

1. Question

1 points

Category: Pediatrics

A neonate is very small for gestational age and shows hypotonia and marked skeletal muscle and subcutaneous fat hypoplasia. During delivery, a large volume of amniotic fluid was released at rupture of membranes. The placenta was small, and only a single umbilical artery was noted. The face has a pinched appearance with hypoplastic orbital ridges, short palpebral fissures, and a small mouth and jaw. The head is small with prominence of the occiput. The ears are low set and malformed. The infant's fists are clenched, with overlapping of the third and fourth fingers. The feet are clubbed, and the great toe is shortened. Which of the following is most likely diagnosis?

1. ☐ 47, XXY
2. ☐ Triple X
3. ☐ Trisomy 13
4. ☒ Trisomy 18
5. ☐ Trisomy 21

INCORRECT ✖

The correct answer is 4.

This is trisomy 18, also known as Edwards's syndrome and trisomy E. The phenotype described in the question stem is typical. Both club feet and rocker bottom feet are common. These infants often have multiple congenital anomalies, which may involve heart, lungs, diaphragm, abdominal wall, or urinary tract. Hernias and cryptorchidism may also occur. Most affected individuals die by 1 year of age; the few survivors usually show marked developmental delay.

(Choice 1), or Klinefelter syndrome, causes males with tall stature and is usually not diagnosed in infancy.

(Choice 2) causes apparently phenotypically normal women and is usually not diagnosed in infancy.

(Choice 3), or Patau syndrome, produces many midline defects, including holoprosencephaly, cleft lip, cleft palate, microphthalmia with colobomas of the irises, scalp defects, and dermal sinuses; other defects are present as well.

(Choice 5), or Down syndrome, is characterized by laterally upward slanting eyes, Brushfield spots of the irises, simian creases, and cardiac anomalies.

2. Question

1 points

Category: Pediatrics

A child was in her usual state of good health when she first developed a focal seizure while playing with her sister. The mother, a registered nurse, states that the seizure lasted approximately 5 minutes. The 911 Emergency Services were called, and the child arrived postictal to the emergency department approximately 1 hour after the seizure first occurred. On physical examination, her vital signs are stable, but the patient is noted to have weakness and hemiparesis. Remarkably, within 24 hours, the weakness and neurologic deficits disappear. Which of the following is the most likely diagnosis?

1. ☐ Hemiplegia complicating a migraine headache
2. ☐ Spastic hemiplegia
3. ☐ Postviral encephalitis
4. ☒ Todd's paralysis
5. ☐ Infratentorial tumor

INCORRECT ❌

The correct answer is 4.

Todd's paralysis commonly occurs after a seizure. The hemiparesis lasts minutes to hours but resolves within 24 hours. It can easily be confused with a stroke except for its duration.

(Choice 1) Hemiplegia complicating migraine headaches also shows recovery within a few hours without seizures, but residual neurologic effects are associated with transient hemiparesis/plegia.

(Choice 5) The disorder usually involves a strong maternal history of classic migraine. Neurologic deficits would not disappear after 24 hours in patients with infratentorial tumor.

(Choice 3) In postviral encephalitis, focal neurologic signs may be stationary, fluctuating, or progressive.

(Choice 2) Spastic hemiplegia occurs in patients with cerebral palsy who have decreased spontaneous movements on one side of their body. These patients may also demonstrate hand preference at a very early age. This problem is not transient.

3. Question

1 points

Category: Pediatrics

An 11-month-old girl is brought by her mother to the office due to fever, fussiness, and irritability. The girl has a tendency to hold and pull on her left ear. For the past few days, she has had rhinorrhea and nasal congestion. Her medical history reveals nothing particular. Her temperature is 38.9 °C (102 °F). Otoscopic examination reveals a bulging, erythematous tympanic membrane with decreased mobility on air compression. What is the most likely organism responsible for the patient's condition?

1. ☐ Haemophilus influenzae
2. ☒ Streptococcus pneumoniae
3. ☐ Moraxella catarrhalis
4. ☐ Pseudomonas aeruginosa
5. ☐ Group A Streptococcus

INCORRECT ❌

The correct answer is 2.

This patient's clinical presentation points to the diagnosis of acute otitis media. The most common causative organism is Streptococcus pneumoniae (40% of cases), followed by non-typable Haemophilus influenzae (25-30%), and Moraxella catarrhalis (10-15%).

Approximately 30% of cases are caused by viruses, especially respiratory syncytial virus and rhinovirus. Approximately 5% of cases are caused by other bacteria (e.g., Group A streptococcus, Staphylococcus aureus, and Pseudomonas aeruginosa). The risk factors of otitis media include age (6-36 months), low socioeconomic status, use of formula milk, caretaker smoking, respiratory allergy, craniofacial anomalies, and chronic middle ear effusion. The complications are numerous, and include chronic suppurative otitis media, acute mastoiditis, labyrinthitis, facial paralysis, cholesteatoma, tympanosclerosis, perforation, conductive hearing loss, and permanent sensorineural hearing loss. The first-line treatment is a ten-day course of amoxicillin. If this regimen fails, amoxicillin clavulanic acid, cefuroxime axetil, and IM ceftriaxone are good alternatives.

(Choice 1) Haemophilus influenza is found in 25-30% of cases, and is the second most common pathogen.

(Choice 3) Moraxella catarrhalis is the third most common causative organism of acute otitis media (found in 10-15% of cases).

(Choice 4) **S. aureus and gram-negative organisms are found most commonly in neonates and very young infants who are hospitalized.**

(Choice 5) Group A Streptococcus, S. aureus and gram-negative bacteria are responsible for 5% of cases of otitis media.

4. Question

1 points

Category: Pediatrics

A neonate is noted to have an abnormally shaped face with a very small jaw. Several hours after birth, the baby develops convulsions and tetany. Serum chemistries show the following:

Sodium 1: 40mEq/L

Potassium: 4 mEq/L

Chloride: 100 mEq/L

Bicarbonate: 24 mEq/L

Magnesium: 2 mEq/L

Calcium: 5 mg/dl

Glucose: 100 mg/dL

This child's disorder is associated with aplasia or hypoplasia of which of the following organs?

- 1. ☐ Ovaries
- 2. ☐ Pancreas
- 3. ☐ Pituitary
- 4. ☒ Thymus
- 5. ☐ Thyroid

INCORRECT ❌

The correct answer is 4.

The only abnormal finding in the serum chemistries is the low calcium level. The disease is DiGeorge syndrome, in which fetal malformations of the third and fourth branchial arches cause failure of development of both the parathyroid glands and the thymus. An abnormal face with a small jaw may be noted at birth. The absence of the parathyroid glands causes hypocalcemic convulsions and tetany in infancy. Aplasia or near aplasia of the thymus predisposes for infections; circulating T cells may be very low to absent. The presence of even a small amount of functioning thymic tissue may allow these children to "outgrow" their immunodeficiency over a period of 4 or 5 years; children with severe disease often die. These children are also likely to have cardiac malformations, which may cause death.

(Choice 1) Abnormal ovaries are associated with Turner syndrome.

(Choice 2) The pancreas is vulnerable to congenital anomalies either as an isolated finding or in association with other congenital anomalies, but the description is most suggestive of DiGeorge syndrome.

(Choice 3) Developmental abnormalities of the pituitary gland are unusual, possibly because significant in utero damage to this important endocrine organ leads to fetal death.

(Choice 5) Developmental abnormalities of the thyroid gland can cause cretinism (infantile hypothyroidism).

5. Question

1 points

Category: Pediatrics

A low-birth-weight infant presents to his pediatrician at 2 months of age. The mother states that the infant has not been eating well. Upon physical examination, the infant is noted to be pale and tachycardic. The lungs are clear to auscultation, and there is no hepatosplenomegaly. A complete blood count shows the hemoglobin level to be 6 g/dL. Which one of the following is the most likely cause of anemia in this infant?

1. ☐ Megaloblastic anemia
2. ☐ Sickle cell anemia
3. ☒ Anemia of prematurity ✓
4. ☐ α -thalassemia
5. ☐ Homozygous β -thalassemia

INCORRECT ✖

The correct answer is 3.

Anemia of prematurity occurs in low-birthweight infants approximately 1–3 months after birth. It is caused by the physiologic effects of the transition from fetal to neonatal life, as well as by factors such as shortened red blood cell survival, rapid growth, and frequent phlebotomy for blood tests. Hemoglobin (Hb) levels in anemia of prematurity are below 7–10 g/dL. Clinical manifestations may include feeding problems, tachypnea, tachycardia, and pallor.

(Choice 1) Megaloblastic anemia of infancy usually is caused by a deficiency of folic acid and has its peak incidence between 4–7 months of age. Infants with folate deficiency are irritable, have poor weight gain, and have chronic diarrhea.

(Choice 2) Newborns with sickle cell anemia rarely exhibit clinical manifestations until 5–6 months of age, when the γ -globin of fetal Hb is replaced by the β -globin of adult Hb. Acute sickle dactylitis (i.e., painful swelling of the hands and feet) is usually the first evidence that sickle cell disease is present in an infant. The thalassemias are a group of heritable, hypochromic anemias.

(Choice 4) is caused by abnormalities in the synthesis of the α -chains of Hb and is manifested as microcytosis of the newborn (mean corpuscular volume [MCV] <95 pg).

(Choice 5) usually manifests as a severe, progressive hemolytic anemia in the second 6 months of life (after Hb A replaces Hb F) and requires regular blood transfusion.

6. Question

1 points

Category: Pediatrics

A 5-year-old girl is brought to medical attention by her parents 12 hours after the onset of generalized tonic-clonic seizures. She never had similar episodes in the past. The girl is otherwise healthy. Her temperature is 37.0 C (98.6 F). Physical examination is unremarkable. There is no evidence that the girl had a fever at the onset of the convulsive episode. The parents fear that seizures may damage the child's brain and may eventually recur. Which of the following is the most appropriate next step in management?



1. ☒ Provide reassurance to child and family ✓
2. ☐ Admit patient to the hospital for further evaluation
3. ☐ Perform electroencephalographic studies
4. ☐ Perform CT/MRI studies of the brain
5. ☐ Start antiepileptic medication

INCORRECT ✗

The correct answer is 1.

After a first seizure, there is a high likelihood (approximately 60 to 70%) that a child will not experience another seizure episode again. Extensive laboratory examination and hospital admission **(Choice 2)** are not necessary, unless physical examination reveals some organic disease that may be related to the onset of the seizure. However, the majority of seizures of new onset are idiopathic, i.e., no cause can be identified with the commonly available diagnostic tools. Children developing a first-time seizure do not need antiepileptic drugs.

Furthermore, approximately two thirds of children who develop a second episode will not have additional seizures. Medication may reduce the risk of having a second or third episode but does not affect the long-term outcome, i.e., the probability that a patient will be seizure-free 2-3 years after the first seizure. Thus, the only measure to take in this situation is to provide reassurance to both patient and family by explaining that the child probably will not have any other seizures and that isolated seizures do not cause any damage to the brain.

(Choice 3) would most likely yield no information. In fact, there is not a single test that can diagnose or rule out epilepsy. In addition, continuous EEG monitoring with or without video-monitoring is very expensive. It is indicated only when seizures are frequent during the day and it is necessary to establish a relationship between neurologic manifestations and abnormal electrical activity.

(Choice 4) CT/MRI studies of the brain are rarely necessary after a single episode, unless there is some clinical evidence suggesting a focal intracranial lesion.

(Choice 5) would be unnecessary following a first episode. In fact, there is still controversy whether it should be started even after a second episode (that is, once a diagnosis of epilepsy is made).

7. Question

1 points

Category: Pediatrics

A 3-year-old girl is brought to the emergency department because she became partially unconscious, lethargic and febrile. Her mother claims that "she suddenly took a turn for the worse" 24 hours ago after an upper respiratory tract infection. She has no allergies, and was not taking oral antibiotics. Her blood pressure is 60/45mm Hg and pulse is 120/min. On examination, she appears emaciated and lethargic. Despite numerous attempts, it is impossible to start a peripheral intravenous line, and the child's condition continues to deteriorate. What is the best next step to obtain IV access in this patient?

1. ☐ Subclavian central line
2. ☒ intraosseous 
3. ☒ Jugular cut down 
4. ☐ Femoral vein
5. ☐ Fluids down endotracheal tube

INCORRECT 

The correct answer is 2.

Whenever intravenous access cannot be obtained in emergent pediatric cases, intraosseous access should be attempted next.

Intraosseous cannulation can be performed quickly in the ED, even with less than optimal light, assistance and choice of equipment.

Another advantage is that this technique does not require the precision required for cannulation of small vessels, yet provides a cannula large enough to deliver adequate fluids. This route can be used in emergencies for 24 to 48 hours, at which point another intravenous route should have been obtained. Associated complications such as osteomyelitis are rare. In adults, the intraosseous route can also be used, but their greater bone density makes it difficult to penetrate the bone.

(Choice 1) Central line placement in children and infants should only be done by highly-skilled individuals,

and is therefore not recommended in the ED. Subclavian line placement in children is associated with a high incidence of complications such as pneumothorax and hemothorax.

(Choice 3) Jugular vein cut down in the ED is not recommended. This is difficult to perform in the absence of adequate lighting and appropriate instruments. Furthermore, the jugular vein area is not the recommended site for cut down as many important structures are found in the neck area.

(Choice 4) Femoral vein cannulation can be done and is a relatively easy technique; however, many do not prefer this technique in children because of possible perineal contamination. Furthermore, lines placed in the groin area make it difficult for patients to be mobile, and bleeding from the site is a constant problem.

(Choice 5) Fluids should never be placed down the endotracheal tube because this leads to aspiration and symptoms of ARDS (Acute Respiratory Distress Syndrome). Only in urgent situations should medications such as epinephrine or lidocaine be placed down the endotracheal tube.

8. Question

Category: Pediatrics

1 points

A 15-month-old Caucasian boy is brought to the office due to fever for the past 3 days. He was born after an uncomplicated pregnancy. His growth and development had been normal until 6 months of age, after which he had 4 episodes of otitis media and two episodes of pneumococcal pneumonia. Physical examination reveals a febrile, irritable boy who is constantly pulling his ears. He has bulging tympanic membranes on the right side, with poor light reflex. His total immunoglobulin level is 40mg/dl. His serum concentrations of IgA, IgM, IgD, and IgG are decreased. The smear reveals an absence of circulating B cells. What is the most likely diagnosis?

1. ☒ Bruton's agammaglobulinemia
2. ☐ Common variable immunodeficiency
3. ☒ Transient hypogammaglobulinemia of infancy
4. ☐ Selective IgA deficiency
5. ☐ DiGeorge syndrome

INCORRECT ❌

The correct answer is 1.

Bruton's agammaglobulinemia, also known as X-linked agammaglobulinemia, is an X-linked recessive disorder characterized by a defect in tyrosine kinase in B cells. This patient's presentation is classic: a male infant who is initially asymptomatic until 6-9 months of age, after which he begins to experience recurrent pyogenic (S. pneumoniae and H. influenzae) infections. The maternal IgG antibodies are responsible for keeping the infant free from infections in the first 6-9 months of life. The diagnosis is confirmed by decreased serum concentrations of IgG, IgA, IgM and IgE, along with absent or markedly decreased B cells on the smear.

(Choice 2) Common variable immunodeficiency (CVID) or acquired hypogammaglobulinemia can have a somewhat similar presentation to Bruton's agammaglobulinemia; however, patients with the former usually manifest with less severe symptoms at a later age (approximately 15- to 35-years-old). The serum concentrations of IgG, IgA, IgM and IgE may also be decreased, but there is no absence or decrease in the number of circulating B cells. Furthermore, there is no sexual predominance in CVID.

(Choice 3) Transient hypogammaglobulinemia (THI) is characterized by decreased IgG levels, normal IgA levels, and variable IgM levels. These immunoglobulin levels all normalize by 6-11 months of age.

(Choice 4) Selective IgA deficiency is characterized by decreased IgA levels (less than 50mg/ml) with normal serum concentrations of other immunoglobulins (although IgG2 subclass deficiency may occur). There is no other evidence of any immune defects. Patients commonly present with respiratory, gastrointestinal, and genitourinary infections.

(Choice 5) The typical patient with DiGeorge syndrome is a neonate with hypocalcemic seizures. The underlying pathology is a defect in the development of the 3rd and 4th pharyngeal pouches, thereby leading to hypoplastic or aplastic thymus and parathyroid glands. Other congenital defects that may occur with this syndrome are esophageal atresia, congenital heart disease, anomalies of the great vessels, mandibular hypoplasia, and low-set, notched ears.

9. Question

1 points

Category: Pediatrics

A 10-day-old infant is brought to the office by her mother for the evaluation of purulent discharge from both eyes. The discharge appeared three days ago, after which the child developed a cough. On examination, the infant's eyes appear normal, except for the presence of purulent discharge. Auscultation reveals diffuse rales without wheezing. Chest radiograph shows a hyperinflated thorax. What is the most likely diagnosis?

1. ☐ Gonococcal conjunctivitis
2. ☐ Chemical conjunctivitis
3. ☒ Congenital chlamydial infection ✓
4. ☐ Congenital rubella infection
5. ☐ Adenovirus infection



INCORRECT ✗

The correct answer is 3.

This newborn most likely has conjunctival and pulmonary chlamydial infection. Chlamydia is usually transmitted from the mother to the neonate during passage through the birth canal. Infected infants have a 25% to 50% risk of developing conjunctivitis, and a 5% to 20% risk of developing pneumonia. Chlamydia is the most common causative agent of infectious neonatal conjunctivitis. Chlamydia/conjunctivitis develops a few days to several weeks after birth, and manifests with conjunctival congestion, edema, and mucoid or frank purulent discharge. Pneumonia usually appears 3 to 19 weeks after birth, and symptoms include cough, tachypnea, and rales; there is no fever, and wheezing is rare. (The latter two features permit to differentiate chlamydial pneumonia from RSV bronchiolitis.) The treatment is oral erythromycin for 14 days, as topical agents are ineffective.

Aside from an increased incidence of neonatal conjunctivitis and pneumonia, cervical carriage of Chlamydia has been associated with a higher incidence of preterm labor, PROM, and late post-term endometritis. All these adverse outcomes and neonatal disease is best prevented by early detection and treatment. Screening is thus recommended in high-risk individuals such as single mothers, adolescents, patients with low socioeconomic status, and promiscuous patients. Erythromycin is the drug of choice in pregnancy and should be given to both the patient and her sexual partner. Another test should be performed later in pregnancy, and a positive result warrants a second course of antibiotics.

(Choice 1) Gonococcal conjunctivitis is extremely purulent and presents between the 2nd and 5th day after birth.

(Choice 2) Chemical conjunctivitis is secondary to the administration of prophylactic drops of silver nitrate. The conjunctivitis is mild and resolves within 24 hours without treatment.

(Choice 4) Ocular disease of congenital rubella syndrome is more serious and may include microphthalmia, cataract, glaucoma and retinopathy.

(Choice 5) The discharge in adenovirus conjunctivitis is also accompanied with respiratory symptoms but of the upper respiratory tract. Moreover, the discharge is usually serous rather than mucoid or purulent, and is associated with mild itching.

10. Question

1 points

Category: Pediatrics

A 2-year-old boy is brought to the emergency department because of increasing and profound lethargy during the past 4 hours. The past medical history is unremarkable. However a 7-year-old brother who is a bed wetter has an upper respiratory infection. There have been no other contacts with individuals who are sick. On physical examination, the child's temperature is 37.4°C (99.3°F), and he is noted to be hypotensive. There is no history or evidence of trauma. The cardiac monitor demonstrates a ventricular dysrhythmia, and the child begins to have a seizure. Which of the following is the most likely cause for this child's problem?

1. ☐ Shaken baby syndrome
2. ☐ Meningitis
3. ☒ Imipramine ✓
4. ☐ Congenital heart disease
5. ☐ Febrile seizure

INCORRECT ✗

The correct answer is 3.

The patient is a toddler and at risk for accidental ingestion. Imipramine is an antidepressant sometimes used to treat nocturnal enuresis. Children who have ingested tricyclic antidepressants primarily present with status epilepticus, coma, and dysrhythmias.

(Choice 1) Seizures may also be associated with shaken baby syndrome, but retinal hemorrhages are usually noted on physical examination.

(Choice 2) This patient is afebrile. Although meningitis should be included in the differential diagnosis of a lethargic child, it is less likely given the fact that the child presents with symptoms associated with tricyclic antidepressant ingestion, and the child's brother is a bed wetter.

(Choice 4) usually manifests itself earlier than 2 years of age.

(Choice 5) are associated with a rapid rise in temperature and usually develop when the core temperature reaches 39.0°C (102.2°F) or above.

11. Question

1 points

Category: Pediatrics

A mother brings her 12-month-old child to the emergency department because he is having painless rectal bleeding. She tells this physician, new to her, that approximately 2 months ago the patient was evaluated for anemia by his primary care pediatrician and was prescribed supplemental iron for presumed iron deficiency anemia. However, the iron deficiency anemia has been refractory to iron therapy. On physical examination, the child's stool is repeatedly positive for occult blood. Which of the following is the most likely diagnosis?

1. ☐ Intussusception
2. ☐ Duodenal ulcer
3. ☐ Adhesions
4. ☒ Meckel's diverticulum
5. ☐ Ulcerative colitis



INCORRECT ❌

The correct answer is 4.

Meckel's diverticulum usually presents as painless rectal bleeding. The condition is best remembered as the disease of twos; it affects 2% of the population, occurs in the first 2 years of life, and is a sacculization 2 feet proximal to the ileocecal junction. Iron deficiency anemia can result from chronic blood loss. The diverticulum usually consists of ectopic gastric tissue. Diagnosis is made by Meckel radionuclide scan, which is performed after intravenous infusion of technetium-99m pertechnetate. Surgical excision is the treatment of choice.

(Choice 1) presents as an acute abdomen with currant-jelly stool.

(Choice 2) Patients with duodenal ulcers have pain.

(Choice 3) Adhesions are fibrous bands that may cause bowel obstruction after abdominal surgery, causing abdominal pain, nausea, and vomiting. They are not associated with rectal bleeding.

(Choice 5) typically presents with bloody diarrhea with mucus.

12. Question

1 points

Category: Pediatrics

A 1-day-old male infant has bilious vomiting after every feeding. He hasn't passed any stools yet. He had no prenatal care, and was delivered vaginally at term. He weighs 3kg (6.61b), and his APGAR scores were 6 and 7 at 1 min and 5 min, respectively. On examination, the neonate is hypotonic, he has a flat facial profile, short ears with down folding ear lobes, a single palmar crease, and a depressed fontanel. There is abdominal distention which is most prominent in the upper abdomen. His abdominal x-rays show gastric and duodenal gas distension with no air distally. What other anomaly can also be expected in this infant?

1. ☐ Sigmoid volvulus
2. ☐ Meconium plug syndrome
3. ☒ Hirschsprung's disease ✓
4. ☐ Intussusception
5. ☐ Meckel's diverticulum

INCORRECT ✖

The correct answer is 3.

The clinical picture is consistent with Down's syndrome, and this newborn is most likely suffering from duodenal atresia. Duodenal atresia is the most common gastrointestinal anomaly in Down's syndrome patients. Typically a "double bubble sign" is seen on abdominal radiograph. Other GI malformations that can occur are:

- Hirschsprung's disease
- Esophageal atresia
- Pyloric stenosis
- Malrotation of the bowel

Congenital heart disease is the most common cause of death in childhood. The common cardiac defects that can occur are:

- Endocardial cushion defect
- Ventricular septal defect
- Patent ductus arteriosus

(Choices 1, 2, 4 & 5) Sigmoid volvulus, meconium plug syndrome, intussusception and Meckel's diverticulum are not associated with Down's syndrome.

13: Question

1 points

Category: Pediatrics

An otherwise healthy 13-year-old boy has seasonal allergic rhinitis. He complains of excessive rhinorrhea, frequent sneezing, and nasal congestion. He has a nasal voice and breathes with his mouth. He derives some relief from keeping windows closed at home and spending as little time as possible outdoors in periods of high pollen concentration. However, he is excessively bothered by nasal congestion. Which of the following drugs would be most effective in relieving nasal congestion?

1. ☒ Alpha-adrenergic agents such as phenylephrine ✓
2. ☐ Antihistamines such as chlorpheniramine
3. ☐ Antihistamines such as loratadine
4. ☐ Cromoglycate or similar mast cell stabilizers
5. ☐ Ipratropium bromide

INCORRECT ✗

The correct answer is 1.

Nasal congestion, rhinorrhea, sneezing, and itchy nose and eyes are the most characteristic manifestations of allergic rhinitis, whether due to seasonal or perennial triggers. Nasal congestion can be particularly bothersome to patients with this condition. It is due to edema of the nasal mucosa, with resultant narrowing of the nasal passages. Patients are forced to breathe through their mouth, causing nasal speech and snoring. Over time, children will acquire what is known as "adenoidal fades," with flat malar eminences, narrow maxillae, and an arched palate. Pharmacotherapy becomes necessary if avoidance of allergens is not feasible or sufficient to provide satisfactory relief. Alpha-adrenergic drugs, such as phenylephrine and pseudoephedrine, are most effective in relieving nasal congestion because of their vasoconstricting action. Two important side effects are rebound congestion and rhinitis medicamentosa.

(Choice 2) Antihistamines such as chlorpheniramine, i.e., first-generation antihistamines, are useful in treating symptoms of rhinitis, especially sneezing, rhinorrhea, and itching, but are less effective in preventing congestion. These drugs cause sedation and anticholinergic side effects.

(Choice 3) Antihistamines such as loratadine, i.e., second-generation antihistamines, do not cause sedation and have therapeutic effects similar to the first-generation antihistamines.

(Choice 4) Cromoglycate, or similar mast cell stabilizers, have therapeutic effects comparable to antihistamines but are not useful in relieving nasal congestion. These drugs are most effective when administered prior to allergen exposure.

(Choice 5) is an anticholinergic agent that blocks cholinergic-mediated mucus secretion and vasodilatation. It can prevent rhinorrhea but is less effective for congestion.

14. Question

1 points

Category: Pediatrics

A 4-week-old male infant has been spitting up his formula feedings for the past few days. He does not vomit bilious material or blood. The spitting up is gradually becoming more frequent, and forceful vomiting ensues. The vomitus seems to shoot straight out and nearly hit the wall. On examination, the baby seems hungry and is chewing his fist. His mucous membranes appear dry. A small, round mass, about the size of an adult thumbnail, is palpated in the upper abdomen.

Laboratory data reveals:

Na^+ : of 133 mEq/L,

K^+ : of 3.5 mEq/L,

Cl^- : of 93 mEq/L

HCO_3^- : of 29 mEq/L.

Which of the following is the most appropriate next step in management?

- ☐ Change the feedings to clear liquids or Pedialyte
- ☒ Obtain a surgical consult immediately ✓
- ☐ Obtain flat plate and upright x-ray films of the abdomen
- ☐ Insert a nasogastric tube
- ☐ Begin parenteral antibiotics

INCORRECT ✗

The correct answer is 2.

This infant has the classic presentation of pyloric stenosis. It occurs more commonly among first-born males, particularly white males. The cause of pyloric stenosis is unknown. It usually presents with nonbilious vomiting at 3-6 weeks of age. It progresses from intermittent vomiting to increasing numbers of episodes with more and more forceful vomiting. The infant acts hungry afterward, but as the vomiting continues there is loss of fluids and electrolytes resulting in hypochloremic metabolic alkalosis. Abdominal examination usually reveals a pyloric lump that feels like an olive-size mass. Treatment for pyloric stenosis is surgical; therefore, immediate consultation with the surgeon is necessary. The surgeon will preoperatively correct the metabolic alkalosis and surgically perform a pyloromyotomy.

(Choice 1) Changing the feedings to clear liquids and a solution, such as Pedialyte, will not relieve this obstruction and will only delay the needed surgical treatment.

(Choice 3) Obtaining abdominal x-ray films will not delineate the specific problem. Appropriate x-ray films in this situation include either a barium swallow, which will reveal a large stomach with only thin streaks of barium in the pylorus due to the hypertrophied pylorus, or an ultrasound, which will reveal a doughnut appearance of the pyloric area.

(Choice 4) will relieve an overdistended stomach, but will not treat the underlying problem.

(Choice 5) Treating the infant with parenteral antibiotics is not indicated. This is not an infectious condition, and prophylactic antibiotics are not used in conjunction with the surgery.

15. Question

1 points

Category: Pediatrics

A 9-year-old girl is seen in the office for unsteady gait and weakness in the lower limbs. On examination she is found to have a wide-based gait with constant shifting of position to maintain her balance. There is decreased vibratory and position sense in the lower extremities, and ankle jerks are absent bilaterally. Her feet are deformed with high plantar arches. MRI of the brain and spinal cord shows marked atrophy of the cervical spinal cord and minimal cerebellar atrophy. Nerve conduction velocity results are within normal limits. EKG shows T-wave inversions in the inferior and lateral chest leads. The mother says that her other child, a 3-year-old boy, is completely normal. He shares toys with his sister and plays well with other children at preschool. He is able to copy squares and crosses but is unable to copy a triangle. Which of the following statements would you advance while discussing the girl's condition with her mother?

- ☐ There is nothing to worry about since it's a non-progressive condition
- ☐ It is most likely a result of spontaneous mutation & the chances of your boy developing the same condition are minimal
- ☐ The EKG abnormalities are most likely due to myocardial ischemia
- ☐ The stability of your boy to copy a triangle worries me since it may be an early manifestation of the same condition your daughter is suffering from
- ☒ I advise you to go for prenatal counseling if you desire any future pregnancies ✓

INCORRECT ✗

The correct answer is 5.

The physical findings and MRI features described in the question classically fit into the description of Friedreich Ataxia (FA). It is an autosomal recessive condition characterized by an excessive number of trinucleotide repeat sequences, resulting in an abnormality of a tocopherol transfer protein. The disorder is progressive with poor prognosis (**Choice 1**). Most patients are wheelchair bound by the age of 25, with death occurring by 30-35 years of age.

(**Choice 3**) FA is associated with necrosis and degeneration of cardiac muscle fibers leading to myocarditis, myocardial fibrosis and cardiomyopathy. Cardiac arrhythmia and congestive heart failure contribute to a significant number of deaths. T-wave inversion in this case is most likely due to myocarditis, and not myocardial infarction. [Remember the differential for T-wave inversion: myocardial infarction, myocarditis, old pericarditis, myocardial contusion and digoxin toxicity].

(**Choice 4**) The milestones of the 3-year-old boy are normal and ability to copy a triangle develops after the age of 5 years. (3 years- copies a cross and circle, 4 years- copies a square and rectangle, 5 years copies a triangle, 6 years- copies a diamond.

(**Choices 2**) In families with one affected child, the subsequent risk of another affected child is 25%, which is significant. Genetic counseling is recommended for prenatal diagnosis for parents with one affected child.

16: Question

1 points

Category: Pediatrics

A 6-year-old female with Down syndrome is brought to the physician for behavioral changes. Over the past few weeks, she has begun refusing to do her usual activities. Her parents also report that she seems dizzy and state that she has developed urinary incontinence. On examination, she is hypotonic but hyperreflexic with a positive Babinski reflex. Her gait is ataxic. Which of the following is the most likely diagnosis?

1. ☐ Alzheimer disease
2. ☒ Atlantoaxial instability
3. ☐ Hypothyroidism
4. ☐ Mental retardation
5. ☐ Spinal cord infarction

INCORRECT ❌

The correct answer is 2.

Atlantoaxial instability is a malformation seen in 10-15% of patients with Down syndrome, and most commonly occurs due to excessive laxity in the posterior transverse ligament, which causes increased mobility between the atlas (C1) and the axis (C2). Fortunately, only 1-2% of Down syndrome patients with atlantoaxial instability are symptomatic. Symptoms usually progress over several weeks and result from compression of the spinal cord. Presenting symptoms include behavioral changes, torticollis, urinary incontinence, and vertebral symptoms such as dizziness, vertigo, and diplopia. On examination, upper motor neuron symptoms such as leg spasticity, hyperreflexia, a positive Babinski sign, and clonus are often present. Patients with Down syndrome are normally hypotonic, and they may remain hypotonic or have increased tone with symptomatic atlantoaxial instability.

Atlantoaxial instability is suspected on physical examination and diagnosed with lateral radiographs of the cervical spine in flexion, extension, and in a neutral position. Open mouth radiographs can also be helpful in visualizing the odontoid. Treatment consists of surgical fusion of the first cervical vertebrae (C1) to the second (C2).

(Choice 1) Although there is a higher prevalence of Alzheimer disease among individuals with Down syndrome, it usually presents around ages 40-50.

(Choice 3) The risk of hypothyroidism is increased in individuals with Down syndrome. These patients present with decreased growth velocity, weight gain, cold intolerance, fatigue, constipation, or declining school performance. Neurologic symptoms can include delayed reflexes, but upper motor neuron signs should not be present.

(Choice 4) Mental retardation is seen in individuals with Down syndrome and can vary in its severity. However, progressive neurologic symptoms are not seen with mental retardation alone.

(Choice 5) Spinal cord infarction presents with the acute onset of severe pain, weakness, and paresthesias. This condition is rare and more commonly seen in adults, and there is no increased risk in patients with Down syndrome.

17. Question

1 points

Category: Pediatrics

The 1-year-old brother of a child with known abetalipoproteinemia is evaluated by a pediatrician for the disease. The 1-year-old has been exhibiting steatorrhea and ataxia. Which of the following would most strongly support the suspected diagnosis?

1. ☒ Acanthocytes on peripheral smear ✓
2. ☐ "Crumpled silk" histiocytes on bone marrow biopsy
3. ☐ Globoid cells on brain biopsy
4. ☐ Metachromatic deposits on sural nerve biopsy
5. ☐ "Sea-blue" histiocytes on bone marrow biopsy

INCORRECT ✗

The correct answer is 1.

➤ Numerous acanthocytes (erythrocytes with spiny membrane projections) on peripheral smear are a feature of abetalipoproteinemia, which is caused by a mutation in the gene for microsomal triglyceride transfer protein. Abetalipoproteinemia is a rare, usually autosomal recessive, congenital disorder characterized by a complete absence of β -lipoproteins, steatorrhea, and blindness due to retinitis pigmentosa, ataxia, and mental retardation. The complete absence of β -lipoprotein means that neither chylomicrons nor VLDL is formed. Massive-dose vitamin A and E therapy has delayed neurologic deterioration in some cases.

(Choice 2) suggest Gaucher disease.

(Choice 3) suggest Krabbe disease.

(Choice 4) suggest metachromatic leukodystrophy.

(Choice 5) suggest Niemann-Pick disease.

17. Question

1 points

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(Choice 5) suggest Niemann-Pick disease.

18. Question

1 points

Category: Pediatrics

A 9-month-old, chubby, healthy-appearing boy is brought to the pediatrician because of episodes of colicky abdominal pain and blood-tinged stools. The pain lasts from 1 to 10 minutes and causes the infant to double up; he then appears normal until his next bout of colic. During the examination, the infant has another episode, at which time a vague mass can be felt on the right side of the abdomen, and the right lower quadrant has an "empty" feeling to deep palpation. Which of the following is the most appropriate initial step in management?

1. ☒ Barium enema ✓
2. ☐ Colonoscopy
3. ☐ Gastrografin enema
4. ☐ Upper gastrointestinal endoscopy
5. ☐ Exploratory surgery

INCORRECT ✗

The correct answer is 1.

The clinical picture is that of intussusception, which occurs in this age group with the presentation given and the classic bloody stools (often referred to as "currant jelly stools") mentioned. A barium enema is both diagnostic and therapeutic, as the heavy column of barium coils around the intussuscepted ileum at the same time that it pushes it back out of the ascending colon.

(Choice 2) could give the diagnosis (in an expensive and invasive way) but would not help resolve the problem. Colonoscopy is extensively used to decompress a distended colon (such as in Ogilvie syndrome), but it offers no way to push back the ileum.

(Choice 3) also has a dual diagnostic/therapeutic role in pediatric surgery, but for a different disease: meconium ileus.

(Choice 4) is not the seat of the problem. The bloody stools may have lured you into this choice, but in this case the blood comes from the engorged, irritated, and battered intussuscepted bowel.

(Choice 5) may eventually be needed to fix a recurring problem, but it is not the first step.

19. Question

1 points

Category: Pediatrics

A 7-year-old boy is brought to the physician because of a persistent mucopurulent nasal discharge for 2 weeks following a common cold. The mother also reports that the child has had frequent cough during the day and occasional temperatures up to 38.0 C (100.5 F). The child does not appear critically ill, but he complains of mild pain in the maxillary region and nasal obstruction. Rhinoscopic examination reveals a rivulet of purulent fluid coming from the inferior meatus. The rest of the physical examination is normal. Which of the following is the most likely diagnosis?

1. ☒ Acute bacterial sinusitis ✓
2. ☐ Acute otitis media
3. ☐ Acute viral rhinitis
4. ☐ Allergic rhinosinusitis
5. ☐ Asthma

INCORRECT ✗

The correct answer is 1.

Acute otitis media and acute sinusitis are among the most common infectious conditions of childhood. Acute sinusitis usually follows a common cold and is due to bacterial infection of the maxillary sinuses. It manifests with persistent mucopurulent discharge, daytime coughing without wheezing, and nasal obstruction. The diagnosis is based on a temporal criterion, with such manifestations present for at least 10 days. The 10-daymark allows differentiation of acute sinusitis from acute viral rhinitis (**Choice 3**), which should resolve within a week or so. If nasal discharge and obstruction persist for more than 30 days, the diagnosis is subacute sinusitis. High fever, swelling of eyelids, and pain on slight pressure over the sinuses indicate a severe sinus infection that needs prompt evaluation and aggressive antibiotic treatment. Most cases of acute sinusitis resolve after antibiotic treatment with amoxicillin or a macrolide, plus topical vasoconstrictors and nasal saline lavage for symptomatic relief.

(**Choice 2**) usually follows an upper respiratory infection. It is characterized by otalgia (especially in older children) and the otoscopic findings of bulging and fullness of the tympanic membrane. Acute otitis media is more frequent between 6 months and 3 years of age.

(**Choice 4**) Allergic rhinosinusitis manifests with chronic clear nasal discharge, without fever or pain. There is usually an accompanying history of allergies.

(**Choice 5**) is characterized by paroxysmal attacks of respiratory distress with tachypnea and wheezing. Cough and increased nasal secretion may be present.

20. Question

1 points

Category: Pediatrics

A 25-year old mother takes her 5-year-old child to see a pediatrician for a rash that started on her trunk and then spread to her face and extremities. According to the mother, the rash occurred in varying stages. It began with crops of small, red papules that progressed to teardrop vesicles on an erythematous base. These vesicles became cloudy, broke open, and then formed scabs. Which of the following is the most common complication of this infection in a normal host?

1. ☐ Reye syndrome
2. ☐ Pneumonia
3. ☐ Thrombocytopenia
4. ☐ Encephalitis
5. ☒ Secondary bacterial skin infection ✓

INCORRECT ✗

The correct answer is 5.

The patient has a presentation that is most consistent with varicella (chickenpox). The most common complication of varicella is secondary bacterial infection of skin lesions.

(Choice 1) The association of Reye syndrome with varicella has become rare since salicylates are no longer prescribed as antipyretics for children.

(Choice 2) is more common in adolescents and adults than in children.

(Choice 3) can result in hemorrhages of skin and mucous membranes and may be associated with transient petechiae. Hemorrhagic vesicles can be seen.

(Choice 4) is the most common central nervous system (CNS) complication of varicella but is far less common than secondary bacterial infection of skin lesions.

21. Question

1 points

Category: Pediatrics

An 8-year-old boy is brought to his physician by his mother, who is worried by the child's frequent episodes of daydreaming, which have apparently resulted in a decline in school performance. The child's psychomotor development appears normal. EEG recording reveals bilateral and symmetric 3 Hz spike-and-wave discharges, which begin and end abruptly on a normal background. Which of the following is the most likely diagnosis?

1. ☒ Absence seizures (petit mal) ✓
2. ☐ Complex partial seizures
3. ☐ Pseudoseizures
4. ☐ Simple partial seizures
5. ☐ Tonic-clonic seizures (grand mal)

INCORRECT ✗

The correct answer is 1.

Children with absence seizures manifest periodic and sudden episodes of impaired consciousness, which are often interpreted by the unaware parent as "daydreaming." These episodes may recur frequently during the day (even hundreds of times) and disturb play and school activities. EEG evidence of 3 Hz spike-and-wave activity during attacks is diagnostic of petit mal. The onset of this type of seizures is between the ages of 4 and 12 years; the seizures often resolve before the age of 20 years.

(Choices 2 & 4) Epilepsy can be divided into partial seizures, if the abnormal activity is localized to a specific part of the brain, or generalized seizures, if the entire brain is involved. In complex partial seizures **(Choice 2)**, there is loss of consciousness associated with focal motor (e.g., convulsions and jerking movements) and/or sensory (e.g., visual, auditory, olfactory, and tactile) symptoms that affect different parts of the body depending on the cortical localization. It may be difficult to differentiate complex partial seizures from absence seizures on clinical grounds alone without EEG studies. Simple partial seizures **(Choice 2)** manifest with motor or somatosensory symptoms but with preservation of consciousness.

(Choice 3) refer to attacks that are superficially similar to tonic-clonic seizures but are related to hysterical conversion or malingering. The differential diagnosis with true seizures is usually easy with the aid of EEG studies and serum prolactin measurement. Serum prolactin increases after true tonic-clonic convulsions but is unchanged following pseudoseizures.

(Choice 5) Tonic-clonic seizures are generalized seizures characterized by a sudden loss of consciousness accompanied by a tonic phase (the patient becomes rigid and falls to the ground) and followed by a clonic phase (convulsive manifestations). Each attack usually lasts 2-3 minutes, after which the patient may regain consciousness or fall asleep. Tongue biting and loss of sphincter control are common during the attack.

22. Question

1 points

Category: Pediatrics

A mother presents to the emergency department with her 2-month-old daughter. The child appears lethargic and dehydrated. The mother reports that the patient has not been feeding well for the past few days. She adds that she recently changed the infant's feedings from formula to cow's milk because the infant does not care for the taste of formula. Which nutrient normally in high concentration in cow's milk could cause the symptoms seen in this infant?



1. ☐ Linoleic acid
2. ☐ Linolenic acid
3. ☐ Calcium
4. ☒ Protein ✓
5. ☐ B-complex vitamins

INCORRECT ✗

The correct answer is 4.

Cow's milk is not recommended for children until they are 1 year of age because the high level of protein causes an increased solute load to the kidneys. It is this overload that led to the lethargic, dehydrated state of the 2-month-old infant. (Choices 1,2,3 & 5) Linoleic acid, linolenic acid, calcium, and B-complex vitamins are not present in high concentrations in cow's milk and would not be expected to cause the symptoms noted in this patient.

23. Question

1 points

Category: Pediatrics

A 12-year-old girl is seen by a pediatrician for a mild case of pneumonia. She is treated with an intramuscular injection of penicillin. About 15 minutes later, she develops extreme itchiness, accompanied by the development of wheals scattered over her chest and extremities. She also begins to wheeze and complain of difficulty breathing. The color of her lips and face remains rosy. Which of the following is the most appropriate first step in management?

1. ☒ Epinephrine injection ✓
2. ☐ IV corticosteroids
3. ☐ Intubation
4. ☐ Oral corticosteroids
5. ☐ No specific therapy is needed

INCORRECT ✗

The correct answer is 1.

The girl is having a systemic anaphylactic reaction to the penicillin. The itchiness and wheals (swollen, erythematous patches of skin) are the result of changes in small cutaneous vessels that favor shift of fluid out of the vascular space. The shortness of breath and wheezing are due to edema and bronchoconstriction of the upper airways. Epinephrine injection (either intramuscularly or subcutaneously) is the most appropriate first step in management, as this will usually quickly reverse the anaphylactic reaction if given promptly.

(Choice 2) and histamine blockers are sometimes given after epinephrine in severe cases of anaphylaxis, especially when the stimulating antigen cannot be immediately removed.

(Choice 3) is not initially indicated, since epinephrine usually reverses the airway edema within a few minutes. If her blood begins to desaturate significantly, as evidenced by pulse oximetry, or if respiratory distress or arrest occurs, intubation would be performed.

(Choice 4) are used either as prophylaxis in situations where anaphylaxis may occur, or sometimes after milder cases of anaphylaxis in which the antigen may not have been completely removed.

(Choice 5) is incorrect, since untreated anaphylactic reactions to penicillin can be fatal.

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24. Question

1 points

Category: Pediatrics

A 3-year-old child presents with recurrent right lower lobe pneumonia. His growth parameters are at the 25th percentile, and he is developmentally appropriate for his age. His past medical history is positive for an ear infection at 18 months of age and gastroenteritis at 2 years of age. He has completed two courses of antibiotic therapy with minimal if any improvement. Which of the following conditions is most likely responsible for this patient's disease?

- ☐ Primary Bor T-cell immunodeficiency disorder
- ☐ Cystic fibrosis
- ☐ Chédiak-Higashi syndrome
- ☐ Congenital lung abnormality
- ☒ Foreign body aspiration 

INCORRECT

The correct answer is 5.

Foreign body aspiration is the most common cause of recurrent pneumonia in an otherwise healthy 3-year-old child. Because of the lung's anatomy, right side aspiration is more common. When a child presents with pneumonia, a history of choking can often be elicited.

(Choice 1) Immunodeficiency disorders are characterized by recurrent infections, usually by opportunistic organisms, and failure to thrive.

(Choice 2) Similarly results in failure to thrive, and malabsorption and clubbing are often seen. In cystic fibrosis, common organisms responsible for pneumonia are *Staphylococcus aureus*, *Pseudomonas aeruginosa*, and *Haemophilus influenzae*. In addition, *Burkholderia cepacia* is an organism that is increasing in frequency in patients with cystic fibrosis and may be associated with rapid pulmonary deterioration and death.

(Choice 3) is autosomal recessive and is associated with giant granules in neutrophils and recurrent infections of the skin, mucous membranes, and the respiratory tract. Moreover, the fact that the patient is healthy indicates that this is not Chédiak Higashi syndrome, in which albinism is a key to diagnosis.

(Choice 4) generally present at a much earlier stage.

25. Question

1 points

Category: Pediatrics

A 2 and a half-year-old child is brought to the office for the evaluation of easy bruising, nosebleeds, and decreased activity over the past week. He had an upper respiratory infection that was treated with an antibiotic 2 weeks ago. On examination, he is well-developed, seems well-nourished, anicteric, and pale. Pertinent findings include some small palpable posterior cervical lymph nodes, sinus tachycardia, a grade I/VI systolic ejection murmur, ecchymoses on his left shoulder and both lower extremities, and petechiae over his extremities and groin. There is no hepatosplenomegaly. The laboratory findings are as follows:

Hemoglobin: 7.9 g/dl

Hematocrit: 24%

Platelet count: 12,000/mm³

WBC: 3,000/mm³

Reticulocyte count: 0.5%

A bone marrow biopsy reveal a markedly hypocellular marrow with decreased megakaryocytes and precursors of the erythroid and myeloid cell lines. What is the most likely diagnosis?

1. ☒ Acquired aplastic anemia ✓
2. ☐ Fanconi's anemia
3. ☐ Diamond-Blackfan anemia
4. ☐ Transient erythroblastopenia
5. ☐ Acute myeloid leukemia

INCORRECT ✗

The correct answer is 1.

Acquired aplastic anemia results from an injury to the bone marrow by radiation, drugs (chemotherapy or antibiotics such as chloramphenicol), insecticides, toxins (benzene, carbon tetrachloride), or infections. Signs and symptoms include pallor, fatigue, weakness, loss of appetite, easy bruising, petechiae, mucosal hemorrhage, and fever. Laboratory evaluation demonstrates a normocytic or macrocytic anemia, leukopenia, reticulocytopenia, and thrombocytopenia. A bone marrow biopsy is essential to make the diagnosis; it typically shows profound hypocellularity with a decrease in all cell lines and fatty infiltration of the marrow.

(Choice 2) Classically, patients with Fanconi's anemia have pancytopenia and characteristic congenital anomalies, such as hyperpigmentation on the trunk, neck and intertriginous areas and/or cafe-au-lait spots, short stature, upper limb abnormalities, hypogonadism, skeletal anomalies, eye or eyelid changes, and renal malformations. Blood counts start to decrease between 4 and 12 years of age, and the initial manifestation is usually thrombocytopenia, followed by neutropenia, then anemia.

(Choice 3) Diamond-Blackfan anemia (OBA), or congenital pure red cell aplasia, presents in the first 3 months of life with pallor and poor feeding. CBC reveals a normocytic or macrocytic anemia with reticulocytopenia. WBC and platelet counts are normal.

(Choice 4) Transient erythroblastopenia of childhood (TEC) is an acquired red cell aplasia which occurs in healthy children between 6 months and 5 years old. There is a gradual onset of symptoms such as pallor and decreased activity. The physical examination is unremarkable except for pallor and tachycardia. The typical laboratory findings are normocytic normochromic anemia, with hemoglobin levels ranging from 3 to 8 g/dL, and an extremely low reticulocyte count.

(Choice 5) Bone marrow infiltration due to leukemia results in pancytopenia by crowding out the normal bone marrow elements. Patients present with lethargy (from anemia), bruising and bleeding (from thrombocytopenia), and unexplained fever. They may also complain of bone pain or present with a limp. CBC shows pancytopenia or anemia, thrombocytopenia and leukocytosis. Acute myeloid leukemia is more common in adults. In this case, the classic presentation after an upper respiratory infection, as well as the absence of other features of leukemia (normal bone marrow), makes acquired aplastic anemia the best answer.

26. Question

1 points

Category: Pediatrics

A 12-year-old girl with mild asthma comes to the office for a health maintenance visit. Her mother states that she is using her albuterol inhaler 2 to 3 times a week and that she has a cough that wakes her up at night about 3 times a month. On physical examination, she has diffuse inspiratory and expiratory wheezes. She has no accessory muscle use. Pulse oximetry shows 95% oxygen saturation on room air. Which of the following is the most appropriate treatment for her at this time?

1. ☒ Albuterol nebulized treatment ✓
2. ☐ Cromolyn sodium nebulized treatment
3. ☐ Oxygen via nasal cannula
4. ☐ IV steroids
5. ☐ Subcutaneous epinephrine

INCORRECT ✗

The correct answer is 1.

This child has mild asthma, requiring an albuterol treatment but no other urgent treatment at this time. Albuterol is used as a "rescue" medicine for someone having an acute attack.

(Choice 2) is used as a daily medicine to control asthma and has no benefit in situations requiring an immediate effect. It is not used in acute attacks.

(Choice 3) is unnecessary at this time since she has an oxygen saturation greater than 92%.

(Choice 4) would not be necessary since the patient is experiencing only mild symptoms. If the patient were thought to need steroid treatment, oral steroids would suffice.

(Choice 5) is rarely needed except in life-threatening situations, such as patients with severe asthma in status asthmaticus.

27. Question

1 points

Category: Pediatrics

A term neonate is healthy at birth and receives routine perinatal care. The infant is discharged from the hospital on day 3. Ten days after delivery, the infant develops severe erythema and edema in both eyelids. There is an associated watery discharge that soon became copious and mucopurulent, with presence of pseudomembranes. Which of the following conditions is this infant most at risk for?

1. ☐ Corneal ulceration
2. ☐ Encephalitis
3. ☒ Pneumonia ✓
4. ☐ Sepsis
5. ☐ Silver toxicity

INCORRECT ✗

The correct answer is 3.

This infant has neonatal chlamydial infection that was acquired during birth from an infected mother. The major manifestation presented in this case is chlamydial ophthalmia, which occurs in about 30 to 40% of infants born to infected mothers. The severity of chlamydial ophthalmia can range from mild conjunctivitis to the severe pattern described in the question stem. Follicles are not present in the conjunctiva; they appear in older children and adults. Chlamydial ophthalmia becomes evident 5 to 14 days after birth. Many of these infants also have nasopharyngeal colonization with Chlamydia, and neonatal pneumonia is common in patients who have clinically developed conjunctivitis. Symptoms of chlamydial pneumonia typically occur within 2 to 19 weeks, with an insidious onset of staccato cough.

(Choice 1) Gonorrheal ophthalmia produces an acute purulent conjunctivitis that appears 2 to 5 days after birth or earlier with premature rupture of membranes. It can rapidly lead to corneal ulceration and perforation if treatment is delayed.

(Choice 2) Serious systemic complications, such as encephalitis, can result from herpes simplex keratoconjunctivitis because these infants have poor immunologic response to the generalized herpes infection.

(Choice 4) Sepsis can be a complication of conjunctivitis, but it is less specific to chlamydial ophthalmia compared with other types of bacterial conjunctivitis. The risk for chlamydial pneumonia is more significant in this patient.

(Choice 5) A chemical conjunctivitis may result from prophylactic silver nitrate use in neonates. It usually appears within 6 to 8 hours after installation and disappears spontaneously within 24 to 48 hours. Silver toxicity is not typical of prophylactic silver nitrate use, and it is unrelated to chlamydial ophthalmia.

28. Question

1 points

Category: Pediatrics

A 6-year-old Caucasian boy is brought to the emergency room by his mother with hemiplegia of acute onset. She states that she found the boy unconscious in his room where she had left him playing several minutes ago, and, he slowly gained consciousness, but could not move his right arm and leg. His past medical history is insignificant. Physical examination reveals right hemiparesis with little sensory abnormalities. No meningeal signs are present. The motor function restored spontaneously during 24-hour observation in the hospital. A CT scan of the head is normal. What is the most probable cause of this patient's problem?

1. ☐ Protein C deficiency
2. ☐ Protein S deficiency
3. ☐ Antithrombin III deficiency
4. ☒ Seizure
5. ☐ Migraine

INCORRECT ❌

The correct answer is 4.

The clinical scenario described is suggestive of Todd's paralysis as the cause of this patient's hemiparesis. Sudden loss of consciousness with following disorientation and slow gain of consciousness is a characteristic description of a seizure, if the convulsive episode was missed. Postictal paralysis (Todd paralysis) may follow generalized as well as focal seizures. The exact pathophysiology of this condition is not well understood, but most probably involves alteration in neuronal electrical activity. The motor deficit usually improves rapidly with restoration of the function within 24 hours. Todd's paralysis usually indicates that a structural abnormality underlying the seizure is present. (Choices 1, 2 & 3) Relation of the hemiparesis to the seizure-like episode and rapid reversibility of the motor deficit makes the other causes, like vascular thrombosis unlikely.

(Choice 5) can produce reversible paralysis, but a history of headaches is typical.

29. Question

1 points

Category: Pediatrics

A 9-month-old child presents for well-child care. The patient's head circumference is noted to be two standard deviations above the norm. In addition, the child also has frontal bossing, translucent skin, and eyes that manifest a "setting sun" sign. A meningocele is also evident on physical examination. A computed tomography (CT) scan of the head would show which of the following?

1. ☐ Type I Chiari malformation
2. ☐ Chronic subdural hematoma
3. ☐ Shaken baby syndrome
4. ☒ Protruding cerebellar tonsils ✓
5. ☐ Leptomenigeal cyst

INCORRECT ✗

The correct answer is 4.

The symptoms described indicate that this patient has type II Arnold-Chiari malformation. In this condition, the hydrocephalus is a noncommunicating one. The fourth ventricle is elongated, there is kinking of the brainstem, and portions of the brainstem and cerebellum are displaced into the cervical spinal canal, causing obstruction of cerebrospinal fluid (CSF) flow. Skull films show a small posterior fossa and widened cervical canal (platybasia). Computed tomography or MRI will show hindbrain abnormalities and cerebellar tonsils protruding downward into the cervical canal. In non-communicating hydrocephalus, the most frequent cause is aqueductal stenosis or the Arnold-Chiari malformation associated with meningocele.

(Choice 1) is not associated with hydrocephalus and usually produces symptoms during adolescence or adult life.

(Choice 2) are characterized by headache, personality change, and sudden loss of consciousness.

(Choice 3) Retinal hemorrhages would be seen in children who are victims of shaken-baby syndrome.

(Choice 5) is a rare and late complication of a linear skull fracture and appears as an expanding pulsatile mass on the surface of the skull.

30. Question

1 points

Category: Pediatrics

A neonate has an obviously abnormal foot. The foot is in a markedly plantar flexed position, with the sole facing the adjacent leg in a position of marked adduction. No other anomalies are noted on physical examination. Which of the following is the most likely diagnosis?



1. ☐ Epispadias
2. ☐ Hypospadias
3. ☐ Talipes calcaneovalgus
4. ☒ Talipes equinovarus ✓
5. ☐ Torticollis

INCORRECT ✖

The correct answer is 4.

This is talipes equinovarus, which is the most common form of clubfoot abnormality. The term talipes is used for any deformity of the foot that involves the talus bone. The term equinus refers to a permanent extension of the foot so that only the ball rests on the ground. The term varus refers to a permanent inversion of the foot, so that only the outer side of the sole touches the ground. Talipes equinovarus is best corrected with repetitive casting of the foot beginning at a young age; this approach slowly brings the foot to a more normal position.

(Choices 1 & 2) Epispadias and hypospadias are urethral abnormalities.

(Choice 3) is associated with a flat and dorsiflexed foot.

(Choice 5) is head tilt, which can be present at birth as a result of neck trauma.

31. Question

1 points

Category: Pediatrics

A 2-year-old boy presents to the pediatrician with a 12-hour history of recurrent and severe paroxysmal colicky pain. The pain is accompanied by straining efforts, loud cries, and vomiting. Abdominal distention and tenderness are present on physical examination, and an oblong mass is palpated in the mid-epigastrium. Bloody mucus is noted on the examiner's finger as it is withdrawn after rectal examination. Which of the following is the most likely diagnosis?

1. ☐ Meckel's diverticulum
2. ☐ Congenital pyloric stenosis
3. ☒ An intussusception ✓
4. ☐ Meconium ileus
5. ☐ Necrotizing enterocolitis

INCORRECT ✗

The correct answer is 3.

Intussusception is the most common cause of intestinal obstruction in children between 3 months and 6 years of age, but decreases in frequency after 3 years. The cause is unknown, but it has been associated with gastroenteritis, Meckel's diverticulum, polyps, and sarcoma. Patients have paroxysmal colicky abdominal pain. A sausage-shaped mass can usually be palpated in the right upper abdomen, and bloody, currant jelly stools are passed. Barium enema reveals a coil-spring sign and may even be therapeutic. However, the use of "air" enemas in the diagnosis and treatment of intussusception has supplanted hydrostatic reduction. If enema reduction is unsuccessful, surgical reduction is necessary.

(Choice 1) Meckel's diverticulum usually presents as painless rectal bleeding, and symptoms usually arise within the first 2 years of life, but it is not uncommon for initial symptoms to be seen in the first decade.

(Choice 2) Pyloric stenosis is characterized by non-bilious projectile vomiting in a 1to 3-month-old.

(Choices 4 & 5) Meconium ileus and necrotizing enterocolitis both present in the neonatal period.

32. Question

1 points

Category: Pediatrics

A 14-year-old previously well patient is diagnosed with clinical sinusitis by his physician. A prescription for antibiotics was made out for the patient and given to his parents to treat this condition. The patient's parents had the prescription filled at the pharmacy. Unfortunately, the teenager, who does not like to take medication, was not always compliant and missed many doses of the antibiotic. Presently, he is being evaluated in the emergency department for fever, confusion, extraocular palsies, and proptosis of the eye. Which of the following is the most likely diagnosis?

1. ☐ Hand-Schüller-Christian disease
2. ☐ Graves disease
3. ☒ Cavernous sinus thrombosis ✓
4. ☐ Acute bacterial conjunctivitis
5. ☐ Temporal lobe abscess

INCORRECT ✗

The correct answer is 3.

Cavernous sinus thrombosis is a complication of sinusitis. Symptoms of cavernous sinus thrombosis include ophthalmoplegia and loss of accommodation. Sinusitis also can be complicated by meningitis, epidural or subdural abscesses, optic neuritis, periorbital and orbital cellulitis and abscess, and osteomyelitis of surrounding bones. Complications develop secondary to local extension of the infection.

(Choice 1) has a variable presentation. It may involve skeletal abnormalities, skin lesions, exophthalmos, hepatosplenomegaly, or pituitary dysfunction. Proptosis is not a feature.

(Choice 2) is a hyperthyroid state characterized by irritability, excitability, crying easily, restlessness, and tremors of the fingers.

(Choice 4) presents with generalized conjunctival hyperemia, edema, mucopurulent exudate, and various degrees of ocular discomfort.

(Choice 5) does not present with proptosis.

33. Question

1 points

Category: Pediatrics

The pediatric intern is called to the nursery to examine a small-for-gestational-age newborn. The mother had one prenatal visit during her entire pregnancy. Physical examination is pertinent for hepatosplenomegaly, jaundice, cataracts, and widespread maculopapular lesions of a reddish-blue color. Diagnostic studies are indicative of hepatitis and thrombocytopenia. The mother most likely developed which one of the following diseases during her pregnancy?

1. ☐ Toxoplasmosis
2. ☐ Rubella
3. ☒ Rubella ✓
4. ☐ Varicella
5. ☐ Syphilis

INCORRECT ✗

The correct answer is 3.

Congenital rubella syndrome occurs after German measles infection during pregnancy and causes a variety of congenital malformations. These manifestations may include hepatosplenomegaly, thrombocytopenia, hepatitis, jaundice, cataracts, widespread maculopapular lesions of reddish blue color (i.e., "blueberry muffin" rash), and neurologic depression.

(Choice 1) Clinical manifestations of congenital toxoplasmosis are chorioretinitis, hepatosplenomegaly, jaundice, convulsions, and cerebral calcifications.

(Choice 2) Rubella, aka, measles during pregnancy is not associated with congenital abnormalities.

(Choice 4) Congenital varicella acquired during the first half of pregnancy may cause limb hypoplasia, microcephaly, seizures, and cataracts in the newborn. If transmitted in the last 3 weeks of pregnancy, varicella can be mild or can involve fever and pneumonia.

(Choice 5) Hepatosplenomegaly and generalized lymphadenopathy may be seen in a newborn with congenital syphilis. In addition, the mucocutaneous lesion of syphilis produce snuffles (a purulent and blood-tinged nasal discharge).

34. Question

1 points

Category: Pediatrics

A previously healthy 12-year-old boy is brought to the physician the day after a nocturnal crisis of difficulty breathing, chest tightness, and cough. He has a history of atopic dermatitis that resolved around 6 years of age. He now has no apparent respiratory distress. His breathing is regular, and his respirations are 12/min. Blood pressure, pulse, and temperature are normal. Chest examination reveals only a few crackles that quickly clear after coughing and mild end-expiratory wheezes. Which of the following is the most appropriate next step in diagnosis?

1. ☐ Arterial blood gas analysis
2. ☐ Bronchial provocation test with histamine or methacholine
3. ☐ Complete blood count
4. ☐ Chest x-ray examination
5. ☒ Spirometry before and after administration of a bronchodilator

INCORRECT ❌

The correct answer is 5.

The chest examination may be normal or nearly normal between attacks in patients with mild asthma. A positive personal or family history of other allergic disorders (such as atopic dermatitis in childhood) is frequently found in those with allergic asthma. To confirm a clinical diagnosis, spirometric tests are particularly useful. These should be performed before and after administration of a short-acting bronchodilator to demonstrate that there is airflow obstruction and that this is promptly reversible. Airflow obstruction is supported by finding a forced expiratory volume in 1 second/forced vital capacity (FEV₁/FVC) ratio of <75%.

(Choice 1) is not informative in the diagnosis of asthma, and the results may even be normal during mild exacerbations.

(Choice 2) may be used if spirometry is not diagnostic and asthma is strongly suspected. It must be performed in a specialized setting.

(Choice 3) may reveal blood eosinophilia, which is frequently present in chronic allergic disorders but would not support the specific diagnosis of asthma.

(Choice 4) is necessary only when other disorders that mimic asthma, such as pulmonary infections or pneumothorax, are likely.

35. Question

1 points

Category: Pediatrics

A 3-year-old boy is admitted for seizure-like activity. He has been a healthy child and has been meeting all development milestones. His immunization schedule is up-to-date. Examination is notable for an erythematous throat and fever. His convulsions require IV administration of a benzodiazepine. Serum analysis reveals a normal white cell count with mild basophilic stippling. The lumbar puncture reveals elevated CSF pressure. Head CT scan is notable for cerebral edema. Which of the following is the next diagnostic step?

- ☐ Antistreptolysin O titer
- ☐ Electroencephalography
- ☒ Protoporphyrin level
- ☐ Rapid slide (Monospot) test
- ☐ Spinal fluid culture

INCORRECT ❌

The correct answer is 3.

In the differential diagnosis of acute cerebral edema with convulsions in a child, ingestion of toxic substances, including lead, must be considered early. Lead toxicity is high on the differential, but a history of pica may not be elicited, and basophilic stippling and lead lines may be absent. A lead level is difficult to obtain quickly, however, a free protoporphyrin level may be obtained expediently and is an indication of lead toxicity. Biochemical evidence of lead toxicity includes elevations of blood lead and signs of interference with hemoglobin synthesis, such as an increase in free erythrocyte protoporphyrin.

(Choice 1) A positive antistreptolysin O level is evidence of an infection with *Streptococcus*. Poststreptococcal glomerulonephritis is a frequent cause of the nephritic syndrome in young children, but there is no evidence of a nephritic syndrome, and antistreptolysin O titer is unlikely to be helpful.

(Choice 2) may be helpful to study seizure activity if the initial work-up is negative. Sharp delta waves on the EEG will be indicative of seizure activity.

(Choice 4) The patient's white count is normal, and the Monospot test, is unlikely to be helpful. Heterophil antibody kits such as the Monospot test is simple and fairly sensitive. This antibody can reach high titers in patients who have infectious mononucleosis and will cause sheep erythrocytes to agglutinate.

(Choice 5) Meningitis must be considered, but the possibility of lead poisoning is more important in this circumstance, given the history and the finding of basophilic stippling. Bacterial meningitis is always high on the differential in such situations, and, if the answer is not obtained promptly, a lumbar puncture for cell count should be done and fluid should be cultured before presumptive antibiotics are started.

36. Question

1 points

Category: Pediatrics

A 5-day-old male term neonate has been passing maroon-colored stools for the past three days, along with frequent episodes of vomiting. He is being breastfed by his mother, who is an asthmatic. His vital signs are within normal limits. The physical examination is unremarkable. Stool examination is positive for RBCs and eosinophils. What is the most likely cause of this patient's findings?

1. ☐ Necrotizing enterocolitis
2. ☐ Viral gastroenteritis
3. ☐ Meckel's diverticulum
4. ☒ Milk protein intolerance ✓
5. ☐ Hirschsprung's disease

INCORRECT ✖

The correct answer is 4.

Milk protein intolerance may present with vomiting and bloody diarrhea. Stools may show RBCs and eosinophils, and there may be a family history of an atopic disorder. It is a hypersensitivity reaction to cow's milk proteins, and therefore usually occurs in babies who are fed with cow's milk; however, it may occur in breastfed babies, as the mother's milk may contain proteins from ingested cow's milk.

(Choice 1) Necrotizing enterocolitis occurs in premature babies. It is an acute condition, and abdominal examination shows distension and tenderness.

(Choice 2) In viral gastroenteritis, the associated diarrhea is usually non-bloody.

(Choice 3) Meckel's diverticulum typically presents as painless melena in a child who is approximately 2 years of age.

(Choice 5) Hirschsprung's disease may present with failure to pass meconium within the first 24 hours of life and/or failure to thrive.

37. Question

1 points

Category: Pediatrics

The mother of a 4-year-old child takes her daughter to a pediatrician because she is "scratching all the time." Physical examination demonstrates multiple areas of excoriation, which are worst on the shoulders, buttocks, and abdomen. In the areas where the scratching has occurred, scattered tiny red punctate lesions are also seen. Careful examination of the clothing reveals small, ovoid, grayish-white structures attached to threads on the seams. Which of the following is the most likely causative agent?

1. ☐ *Ancylostoma braziliense*
2. ☐ *Corynebacterium minutissimum*
3. ☒ *Pediculus human us corporis* ✓
4. ☐ *Sarcoptes scabiei*
5. ☐ *Trichophyton rubrum*

INCORRECT ✗

The correct answer is 3.

The child has body lice, *Pediculus humanus corporis*, which live in the clothing and cause itchy bites. The structures on the clothing seams are the eggs. The lice themselves can also often be seen. The lice are confined to the skin surface, and, unlike scabies mites, do not burrow into the skin. The patient, family members, and social contacts (such as other daycare children) should all be treated at the same time with permethrin cream, 5%. Clothing and bedding should also be washed.

(Choice 1) causes cutaneous larva migrans, which produces long itchy burrows on the legs and buttocks.

(Choice 2) causes erythrasma, characterized by pink to brown itchy patches in moist areas of skin.

(Choice 4) causes scabies, which is diagnosed by finding fine white lines up to 1 cm in length (burrows) in the skin, with a mite at the end of them.

(Choice 5) is a fungus that can cause ringworm of the body.

38. Question

1 points

Category: Pediatrics

A 9-month-old child presents for a well-care examination. The mother states that the child has not been feeding well and seems to be less active than usual. The patient's vital signs are within normal limits, and the patient is afebrile. On physical examination, the head circumference is found to be two standard deviations above the norm. The child has frontal bossing, the skin is translucent, and the eyes manifest a "setting sun" sign. The child also has a meningocele. A computed tomography (CT) scan of the head would show which of the following?

1. ☐ Epidural hematoma
2. ☐ Chronic subdural hematoma
3. ☐ Shaken baby syndrome
4. ☒ Noncommunicating hydrocephalus
5. ☐ Leptomenigeal cyst

INCORRECT ❌

The correct answer is 4.

The Arnold-Chiari syndrome type II consists of progressive hydrocephalus and myelomeningocele. The hydrocephalus is a noncommunicating one. The fourth ventricle is elongated, there is kinking of the brainstem, and portions of the brainstem and cerebellum are displaced into the cervical spinal canal, causing obstruction of flow of cerebrospinal fluid (CSF). Skull films show a small posterior fossa and widened cervical canal (platybasia). Type I Chiari malformation is usually not associated with hydrocephalus and is first noted during adolescence or adult life.

(Choice 1) Classically, patients with epidural hematoma experience a brief period of unconsciousness followed by a variable lucid interval and are unconsciousness thereafter. Expansion of the hematoma also leads to headache, vomiting, and focal neurologic signs. Children who are victims of shaken baby syndrome have acute subdural hematomas.

(Choice 2) Chronic subdural hematomas are characterized by headache, personality change, and loss of consciousness.

(Choice 3) In addition, retinal hemorrhages are present, and there may be additional clinical evidence to support child abuse.

(Choice 5) A leptomeningeal cyst is a rare and late complication of a linear skull fracture and appears as an expanding pulsatile mass on the surface of the skull.

39. Question

1 points

Category: Pediatrics

A one-day-old infant with Down syndrome has developed persistent vomiting. He was delivered vaginally at 34 weeks without any complications. On examination, he appears dehydrated and slightly tachypneic. His abdomen is soft and not distended. Abdominal x-ray reveals two large distinct air bubbles, but there are no dilated bowel loops or air fluid levels. What is the most likely diagnosis of this patient?

1. ☐ Pyloric stenosis
2. ☐ Reflux disorder
3. ☐ Tracheoesophageal fistula
4. ☐ Bowel obstruction
5. ☒ Duodenal atresia ✓

INCORRECT ✗

The correct answer is 5.

The above case is a classic example of duodenal atresia. Suspect duodenal atresia when the patient has bile-stained vomitus, no abdominal distension, dehydration and Down syndrome. Vomiting usually begins in the first 24 to 48 hours after birth. The mother may have a history of polyhydramnios. The typical abdominal x-ray finding is the "double bubble" sign (dilated stomach and duodenum). The treatment is decompression with a nasogastric tube, correction of electrolyte abnormalities and surgery.

(Choice 1) Pyloric stenosis is most common in first-born children, and patients may have a positive family history. Features include non-bilious projectile vomiting after feeding, palpable olive-shaped abdominal mass, and visible gastric peristalsis. Hypochloremic metabolic alkalosis is typical. The diagnosis is confirmed with ultrasonography.

(Choice 2) Gastroesophageal reflux is common in infants, and usually occurs in a thriving baby. The vomiting episodes are seen after feeding, and the vomitus is not bilious and rarely contains blood. The infant may present with recurrent pneumonias or cough. A 24-hour pH probe, contrast swallow study, or endoscopy may reveal the diagnosis.

(Choice 3) Tracheoesophageal fistula (TEF) usually presents in infancy with coughing, vomiting and gagging after feeding. Respiratory distress is common after feeding, and frothy bubbles are seen in the oral cavity even after suctioning. The condition may be associated with VACTERL syndromes, and the mother may have a history of polyhydramnios.

(Choice 4) Although malrotation or atresia of the bowel may also present with recurrent vomiting and bile-stained vomitus, the abdomen is usually distended and the typical x-ray finding includes loops of dilated bowel with air fluid levels.

40. Question

1 points

Category: Pediatrics

A 6-year-old boy has multiple, honey-colored, crusted lesions on his face, perioral areas, and forearms. The first lesion appeared 2 weeks ago on his philtrum. Since then the lesions have spread to his hands and arms. Each began as a small pustule on an erythematous base and eventually ruptured to form the crusted lesions now present. His temperature is 100.6 F, pulse is 100/min, and respirations are 14/min. The remainder of the physical examination is unremarkable. Which of the following is the most appropriate treatment?

1. ☐ Clarithromycin
2. ☒ Dicloxacillin ✓
3. ☐ Penicillin G
4. ☐ Penicillin V
5. ☐ Vancomycin

INCORRECT ✗

The correct answer is 2.

This is a case of impetigo, a common superficial infection caused by group A beta hemolytic streptococci or *Staphylococcus aureus*. Dicloxacillin is an oral preparation that is effective for infections caused by staphylococci and some streptococci. The drug is not deactivated by penicillinase, which is often present in staphylococci.

(Choice 1) is a macrolide antibiotic that is used for mild to moderate respiratory tract infections, uncomplicated skin infections, and prophylaxis for *Mycobacterium avium* complex.

(Choice 3) is available for oral, intramuscular, or intravenous use, and is active against streptococci. However, most staphylococci produce β -lactamases, which break down penicillin.

(Choice 4) is an oral preparation of penicillin and has a similar coverage as penicillin G.

(Choice 5) is active against gram-positive species and is reserved for those bacteria that are resistant to β -lactamase-resistant antibiotics. It is used intravenously.

41. Question

1 points

Category: Pediatrics

A 4-day-old, 1,500-g (3 lb 5 oz) premature infant recovering from respiratory distress syndrome is noted to have bounding peripheral pulsations and a hyperactive precordium. A continuous machinery murmur is most audible at the left infraclavicular area. Left ventricle hypertrophy (LVH) is present on the electrocardiogram (ECG). The chest x-ray film shows slight enlargement of the heart and increased pulmonary venous markings. Which of the following is the most likely diagnosis?

1. ☐ Ventricular septal defect (VSD)
2. ☐ Atrial septal defect (ASD)
3. ☒ Patent ductus arteriosus ✓
4. ☐ Pulmonic stenosis
5. ☐ Tetralogy of Fallot

INCORRECT ✖

The correct answer is 3.

A patient with patent ductus arteriosus has a continuous machinery murmur. Bounding pulses with hyperactive precordium also may be found on physical examination. Although the electrocardiogram (ECG) is usually not helpful, it may show left ventricular hypertrophy (LVH).

(Choice 1) Patients with ventricular septal defect (VSD) have a pansystolic murmur.

(Choice 2) Patients with atrial septal defect (ASD) have a widely split and fixed second heart sound.

(Choice 4) In patients with pulmonic stenosis, there is a high-pitched systolic ejection murmur, most audible at the upper left sternal border, that transmits fairly well to the back. A systolic thrill may be present at the upper left sternal border and, rarely, in the suprasternal notch.

(Choice 5) Tetralogy of Fallot is characterized by the presence of a VSD, right ventricular hypertrophy, pulmonic stenosis, and overriding aorta. The murmur of tetralogy of Fallot is a long, systolic ejection murmur most audible at the mid-left sternal border.

42. Question

1 points

Category: Pediatrics

A 13-year-old girl presents with a 1-week history of a sore throat and a nonproductive cough. She has been previously healthy and has not been exposed to any other sick person. She has not been taking any medications. On examination, she has normal oxygen saturation and a low-grade fever. The remainder of the examination is unremarkable. Which of the following is the most appropriate pharmacotherapy?

1. ☐ Amoxicillin
2. ☐ Cefazolin
3. ☒ Erythromycin ✓
4. ☐ Metronidazole
5. ☐ Trimethoprim-sulfamethoxazole

INCORRECT ✗

The correct answer is 3.

The patient is older than 6 years, and the most common pneumonias in this age group are due to *Mycoplasma pneumoniae* and *Streptococcus pneumoniae*. The girl in this vignette most likely has *Mycoplasma pneumoniae*, which is milder than that due to traditional bacteria. It is characterized by gradual onset of fever, headache, and a nonproductive cough. The treatment of choice is erythromycin.

(Choice 1) would be good coverage for *Streptococcus*, but not for *Mycoplasma*.

(Choice 2) would be a good choice for typical bacteria and for coverage against gram-negative rods, but would not be appropriate for *M. pneumoniae*.

(Choice 4) would provide anaerobic coverage, but would not be appropriate for *M. pneumoniae*.

(Choice 5) is good prophylaxis for *Pneumocystis carinii* pneumonia, but this is very unlikely in this presumably immunocompetent patient.

43. Question

1 points

Category: Pediatrics

A 4-day-old female infant presents to the emergency department with vomiting and abdominal distention. The mother states that the vomitus was green. The infant also has had difficulty feeding and has been hard to console. The mother had an uncomplicated pregnancy. The infant passed meconium within 12 hours after birth. She also had several small, seedy, yellowish stools each day since birth. On physical examination, she is very irritable. Her anterior fontanelle is slightly depressed. Her abdomen is distended. Which of the following is the most likely diagnosis?

1. ☐ Allergic reaction to formula
2. ☐ Gastroesophageal reflux disease
3. ☐ Hirschsprung disease
4. ☐ Meconium ileus
5. ☒ Midgut volvulus ✓

INCORRECT ✗

The correct answer is 5.

In the neonatal period, midgut volvulus is the most common cause of abdominal obstruction due to malrotation. Green-colored vomitus represents bilious vomiting, which is caused by obstruction of the small bowel. The infant typically has normal feeding and appears to be well in the first few days of life. As the malrotation worsens, the infant starts to develop abdominal fullness, especially in the right upper quadrant. Bilious vomiting soon develops as the small bowel is completely obstructed. If the volvulus is not diagnosed and treated early, intestinal ischemia and necrosis may develop. This can lead to bowel perforation and shock. Volvulus is the most common type of malrotation in newborns. It is caused by failure of the cecum to move into the right lower quadrant. The usual location of the cecum is in the subhepatic area. Because the cecum fails to rotate properly, it does not form the normal broad based adherence to the posterior abdominal wall. A midgut volvulus is formed when the mesentery, which includes the superior mesenteric artery, is tethered by a narrow stalk and is twisted around itself. In addition, bands of adhesive tissue (Ladd bands) may extend from the cecum to the right upper quadrant, obstructing the duodenum. Midgut volvulus is an emergency in neonates. When an infant is suspected of having this condition, he or she should be stabilized immediately by IV fluids, antibiotics, and nasogastric suctioning. Abdominal radiographs should be taken as soon as possible to evaluate for bowel obstruction. Radiologic findings of volvulus include small bowel dilatation, paucity of air in the intestine, and a corkscrew-like appearance of the duodenum. Emergent surgery is needed to relieve the obstruction. Adhesive tissues should be resected, and the entire intestine should be inspected for anomalies. If segments of ischemic bowel are present, they should be removed. If the viability of the bowel cannot be determined during the surgery, a second surgery 12-36 hours later may be necessary to inspect and remove any ischemic bowel segment. Short bowel syndrome is a dreaded complication if a significant portion of the bowel was nonviable, and thus removed.

(Choice 1) typically presents between 4 and 6 weeks of age. Reactions rarely present in the neonatal period. Bilious vomiting is also not consistent with an allergic reaction to formula.

(Choice 2) happens commonly in infants. In fact, the lower gastroesophageal sphincter is poorly developed during infancy. Most cases of reflux are a normal variant, and the parents need not be worried. However, when reflux causes significant problems, such as poor weight gain or apnea, it then needs to be treated. The vomiting in gastroesophageal reflux disease is never bilious.

(Choice 3) results from partial or complete absence of ganglion cells in the colon. It is the most common cause of neonatal obstruction of the colon. Hirschsprung disease is suspected when the infant fails to pass meconium in the first 24 hours of life. Diagnosis becomes more likely when barium enema shows the appearance of a megacolon proximal to the aganglionic segment. It is confirmed with punch biopsy.

(Choice 4) is most commonly associated with cystic fibrosis. It typically presents at birth or within the first 48 hours. Symptoms include failure to pass meconium, abdominal distention, and vomiting, which may be bilious.

44. Question

1 points

Category: Pediatrics

A 2-year-old child is brought to the emergency department because of generalized convulsions that last 15 minutes. He has had a fever for 24 hours, and his current temperature is 39.5 C (103.0 F). He also has a sore throat, but otherwise looks healthy. His father also had several episodes of febrile seizures in his childhood. Which of the following is the most important factor that will increase the risk of recurrence of febrile seizures?

1. ☐ Age older than 18 months
2. ☐ Duration of seizure longer than 5 minutes
3. ☒ Family history of febrile seizures ✓
4. ☐ Fever of long duration before onset of seizure
5. ☐ Temperature higher than 39.0 C

INCORRECT ✗

The correct answer is 3.

Febrile seizures are common events. This form of benign manifestation affects approximately 3 to 4% of children. Since epilepsy is defined as at least two seizures occurring without an identifiable trigger, febrile seizures do not constitute epilepsy even if they recur. Fever, in fact, is the identifiable trigger. Most children who have an episode of febrile seizure will not have a recurrence. Risk factors that do increase the likelihood of recurrence include family history, age younger than 18 months (**Choice 1**), and temperature less than 39.0 C (**Choice 5**) when seizure occurred. Overall, children with a first febrile seizure will have a recurrence in 25% of cases by 1 year and in 30% by 2 years.

(**Choice 2**) The duration of the first seizure will not influence the likelihood of recurrence, but recurrences will likely also be prolonged if the first episode lasted for more than 15 minutes.

(**Choice 4**) Fever of long duration is negatively correlated to the probability of recurrences.

45. Question

1 points

Category: Pediatrics

A 6-year-old boy is brought to the emergency department because of the acute onset of headache, nausea, and vomiting. On arrival, physical examination reveals marked nuchal rigidity and fundoscopic evidence of papilledema. A head CT scan reveals a solid tumor in the posterior fossa, centered in the cerebellar vermis and extending to the fourth ventricle. An emergency craniotomy is performed, during which a small sample of the tumor is sent to the pathologist for a frozen section consultation. Which of the following is the most likely diagnosis?

1. ☐ Ependymoma
2. ☐ Glioblastoma multiforme (GBM)
3. ☐ Hemangioblastoma
4. ☒ Medulloblastoma
5. ☐ Oligodendroglioma

INCORRECT ❌

The correct answer is 4.

The most common posterior fossa tumors in children include medulloblastoma, ependymoma, and pilocytic astrocytoma.

Medulloblastoma, an anaplastic neoplasm thought to derive from primitive neuroectodermal precursors, belongs to the group of primitive neuroectodermal tumors (PNET). It originates from the cerebellar vermis and grows into the fourth ventricle, often producing signs and symptoms of increased intracranial pressure. The histologic features are those of other small round cell tumors.

(Choice 1) usually derives from the ependymal cells lining the central canal. Childhood ependymomas typically develop in the fourth ventricle and may present with increased intracranial pressure. In contrast to medulloblastoma, however, ependymoma appears on CT/MRI as a mass that fills the fourth ventricle and secondarily involves adjacent structures.

(Choice 2) is a rare tumor in children of this age, and the cerebellum is a rare location at any age. Furthermore, GBM grows in a diffuse fashion within the white matter without involving the ventricular cavities.

(Choice 3) is a vascular tumor of uncertain histologic origin. The cerebellar hemisphere is its most common location, and it often appears as a cystic mass with a mural nodule. Its association with von Hippel-Lindau syndrome is noteworthy.

(Choice 5) is a slow-growing tumor of adults that usually develops in the white matter of a cerebral hemisphere. Calcifications within the tumor are frequently detected on x-ray films or CT scan, and seizures are often the presenting sign.

46. Question

1 points

Category: Pediatrics

A new mother complains that her 6-week-old infant frequently regurgitates small volumes of formula during and after feedings. Physical examination demonstrates a happy baby who has gained half a pound since his last visit. No abdominal masses are noted. Which of the following is the best next step in management?

1. ☐ Change the baby's formula
2. ☒ Change the bottle's nipple 
3. ☐ Monitor the baby carefully
4. ☐ Order abdominal x-rays
5. ☐ Order CT of the abdomen

INCORRECT 

The correct answer is 2.

Regurgitation of small volumes (usually less than 5-10 mL) is common among babies and is usually of no clinical significance unless the baby is failing to grow normally. It is of concern to some mothers, and switching to a firmer nipple bottle with a smaller hole will slow the rate that the baby is drinking and often reduce the amount of swallowed air.

(Choice 1) does not usually alter the amount of regurgitation.

(Choices 3,4 & 5) Careful monitoring of the baby, abdominal x-rays, and CT scan are not warranted for this benign condition.

47. Question

1 points

Category: Pediatrics

A previously healthy 5-year-old girl is brought to the emergency room from a camping trip because her parents are concerned about rabies. This morning, they found a bat in the child's tent. There were no obvious bite marks on the child. Afterwards, the girl helped her father clean animals he had brought back from a hunt, including squirrels, rabbits, and chipmunks. Later, while packing up the campsite, the child was bitten on the arm by a rat. Her encounter with which of the following animals most warrants post-exposure rabies prophylaxis?

1. ☐ Squirrel
2. ☐ Rabbit
3. ☒ Bat ✓
4. ☐ Rat
5. ☐ Chipmunk

INCORRECT ✗

The correct answer is 3.

Rabies is a fatal disease that is transmitted to humans by contact with infected animals. Transmission of the rabies virus requires exposure to saliva or nervous tissue from an infected animal through a bite, open wound, or mucus membrane contact. The need for postexposure prophylaxis is based in part on the animal involved and geographic locale. Bats are a major reservoir for the rabies virus. Bat bites are often so quick and minor that the victim is not even aware. For this reason, any direct contact between bats and humans requires post-exposure rabies prophylaxis. People bitten by domestic animals suspected of being rabid or not available for observation, or by wild carnivores like raccoons, skunks, and foxes should also receive prophylaxis.

(Choices 1,2,4 & 5) Rabbits and small rodents like squirrels, chipmunks, and rats are rarely infected with rabies. Post-exposure prophylaxis following bites is not routine.

48. Question

1 points

Category: Pediatrics

A 12-year-old girl complains of intermittent palpitations. She had previously been in excellent health and has met all development milestones. There is no family history of heart disease. She is on no medications and takes no drugs. She states that the palpitations begin and end suddenly and usually last a couple of hours. She is otherwise asymptomatic between episodes. The physical examination is normal. An ECG reveals a shortened PR interval and a slow upstroke of the QRS wave in lead III. Which of the following is the most likely diagnosis?

1. ☐ Anxiety attack
2. ☐ Lown-Ganong-Levine syndrome
3. ☐ Nodal reentrant tachycardia
4. ☐ Sinus tachycardia
5. ☒ Wolff-Parkinson-White syndrome

INCORRECT ❌

The correct answer is 5.

The shortened PR interval and delta waves with the slow QRS upstroke are classic ECG manifestations of Wolff-Parkinson-White (WPW) syndrome. WPW syndrome is caused by the presence of a tract bypassing the AV node, causing preexcitation and leading to paroxysmal tachycardia. Ablation of the bypass tract is the treatment of choice.

(Choice 1) would present with symptoms of agitation, clamminess, and chest tightness.

(Choice 2) is similar to WPW syndrome, but the PR interval is not shortened.

(Choice 3) would present with P waves following QRS waves.

(Choice 4) would not present with the shortened PR interval and QRS waves.

49. Question

1 points

Category: Pediatrics

An 18-month-old child is brought to the emergency department by his mother due to a one-day history of lethargy and anorexia. He had a fever the whole day yesterday, which responded to Tylenol (Acetaminophen). He then developed a petechial rash over his entire body, which worsened in the last few hours. He is up-to-date with his immunizations, and is an otherwise healthy baby. On examination, he is drowsy and lethargic. He has neck stiffness and appears septic. He flexes his hips when his neck is flexed. What is the most likely organism responsible for the patient's symptoms?

1. ☒ Meningococcus ✓
2. ☐ Haemophilus influenza
3. ☐ Cytomegalo virus
4. ☐ Borrelia burgdorferi
5. ☐ Listeria monocytogenes

INCORRECT ✗

The correct answer is 1.

Suspect meningococccemia in a neonate with signs of meningitis and a petechial rash. Seventy-five percent of patients with Meningococcus meningitis present with a petechial rash that is prominent on the axilla, wrists, flanks and ankles. It appears within 24 hours of the infection, and the patient generally appears sick.

(Choice 2) Haemophilus influenza generally affects 6- to 48-month-old children. It may cause meningitis, but without the rash. It may also cause rhinorrhea, fever, epiglottitis, and ear infections. The antigen is easily detected in the serum and CSF.

(Choice 3) Cytomegalovirus can cause a mononucleosis type picture with high fevers, fatigue, malaise, splenomegaly and pharyngitis. It usually does not cause meningitis, nor does it present with a rash. immunocompromised individuals have a poor prognosis.

(Choice 4) Borrelia burgdorferi causes Lyme disease. It is usually acquired in the late summer months after an Ixodes sp. tick bite. It typically presents with skin rash (i.e., erythema chronicum migrans), headaches, fevers, chills and malaise. The skin rash is very characteristic (i.e., well-circumscribed borders that gets larger in size with time). The disease is also seen in adults, and can cause meningitis.

(Choice 5) Listeria can cause meningitis in the newborn when it is transmitted vaginally, but it does not present with a rash. Another mode of transmission is ingestion of unpasteurized milk or cheese from infected cows. Its incidence is increased in immunosuppressed and diabetic patients.

50. Question

1 points

Category: Pediatrics

A 12-year-oldboy presents to his pediatrician with frequent episodes of headache, nausea, blurry vision, and sweating. On physical examination, his temperature is 37.4 C (99.3F), blood pressure is 148/94mm Hg, pulse is 92/min, and respirations are 18/min. The rest of his examination is unremarkable. His 24-hour urinary vanillylmandelic acid (VMA) and metanephrines are increased. An abdominal CT reveals an extrarenal mass above the left kidney. Which of the following is the most appropriate pharmacotherapy?

- ☒ Alpha-adrenergic blocker
- ☐ Angiotensin-converting enzyme inhibitor
- ☐ Beta-adrenergic blocker
- ☐ Calcium channel blocker
- ☐ Diuretics

INCORRECT

The correct answer is 1.

The clinical presentation of this patient is consistent with pheochromocytoma; this impression is confirmed by the increased levels of urinary catecholamine and metabolites, and an extrarenal mass on abdominal CT. Pheochromocytoma arises from the chromaffin cells in the adrenal medulla or the abdominal sympathetic chain. Clinical manifestations include headache, sweating, nausea, vomiting, palpitation, blurry vision, nervousness, and hypertension. In fact, pheochromocytoma is one of the important causes of secondary hypertension. In a 12-year-oldboy with a blood pressure of 148/94 mm Hg, the differential diagnosis of secondary hypertension should be triggered in every clinician's mind. If pheochromocytoma is suspected, a 24-hour collection of urine for catecholamines and metabolites (vanillylmandelic acid and metanephrines) can be checked. If these substances are elevated, the diagnosis of pheochromocytoma is confirmed. CT scan of the abdomen may be done to locate the tumor. Most Pheochromocytomas are benign. When hypertension is present, as in this clinical vignette, it should be treated promptly, even before surgical resection of the tumor. An α -adrenergic blocker, such as phenoxybenzamine, is the drug of choice because it has been shown to effectively decrease blood pressure in patients with pheochromocytoma.

(Choice 2) does not play any role in controlling blood pressure in children with pheochromocytoma.

(Choice 3) is actually contraindicated in the initial treatment of hypertension in pheochromocytoma because it causes an unopposed α effect that would result in a paradoxical rise of blood pressure. However, labetalol, which is a combined α - and β -blocker, can be used in some patients with efficacy similar to phenoxybenzamine.

(Choices 4 & 5) Calcium channel blockers and diuretics are not used for treatment of hypertension in patients with pheochromocytoma.

51. Question

1 points

Category: Pediatrics

A 2-year-old girl is taken to a pediatrician because she has developed a rash and seems unusually unsteady when she tries to walk. Physical examination demonstrates a diffuse rash on body parts exposed to sun. Also noted are short stature, possible mental retardation, and ataxia. Screening studies demonstrate increased total amino acids in the urine. Which of the following is the most likely diagnosis?

1. ☐ Alkaptonuria
2. ☐ Cystinuria
3. ☒ Hartnup disease ✓
4. ☐ Fanconi syndrome
5. ☐ Phenylketonuria

INCORRECT ✗

The correct answer is 3.

This is Hartnup disease, which is an autosomal recessive condition that produces a neutral aminoaciduria with increased renal clearance of alanine, asparagine, glutamine, histidine, isoleucine, leucine, phenylalanine, serine, threonine, tyrosine, valine, and tryptophan. This is accompanied by malabsorption of other amino acids, notably tryptophan, but also phenylalanine and methionine. The resulting tryptophan deficiency produces pellagra-like symptoms (tryptophan is used to synthesize nicotinamide) with photosensitive skin lesions, ataxia, and neuropsychiatric disturbances. Hartnup disease can be effectively treated (early diagnosis is important to limit neurologic manifestations) with good nutrition supplemented with nicotinamide.

(Choice 1) leads to urinary secretion of homogenetic acid (causing urine that turns black on standing) and causes arthritis and dark coloration of cartilage.

(Choice 2) principally produces urinary tract calculi.

(Choice 4) produces a generalized dysfunction of the proximal tubules with glucosuria, generalized aminoaciduria, and hypophosphatemia (with bony abnormalities).

(Choice 5) Phenylketonuria, which is characterized by excess phenylalanine in urine, is an important cause of mental retardation in young children. Most affected children have pale skin, blond hair, and blue eyes.

52. Question

1 points

Category: Pediatrics

An 8-year-old boy presents to the pediatrician's office with a headache for the past 3 weeks. His mother also states that he has been more tired and has had frequent nose bleeding for the past month. On physical examination, his height and weight are both below the 5th percentile for his age. His blood pressure is 152/86mm Hg in all four extremities. His pulse is 74/min, and respirations are 16/min. His heart examination is normal with no murmur. His peripheral pulses are strong and symmetric. Urinalysis and serum electrolytes are ordered. Which of the following is the most appropriate next step in diagnosis?

1. ☐ 24-hour urine creatinine and protein
2. ☒ Blood urea nitrogen and creatinine concentration ✓
3. ☐ Plasma and urine catecholamine levels
4. ☐ Serum aldosterone level
5. ☐ Serum cortisol level

INCORRECT ✗

The correct answer is 2.

Hypertension happens in children. It is defined as blood pressure higher than the 95th percentile for age on three different measurements. Unlike adults, most cases of hypertension in infants and younger children are secondary hypertension. Children with hypertension usually present with headaches, visual symptoms, easy fatigability, or epistaxis. One of the possible reasons for failure to thrive is hypertension. Seventy to seventy-five percent of children who have hypertension have a renal etiology. Up to 25% of patients have a history of recurrent urinary tract infection secondary to obstructive uropathy or reflux nephropathy. Other renal causes include renovascular disease, glomerulonephritis, and hemolytic uremic syndrome. These children develop hypertension when azotemia occurs because of the underlying disease. Hence, the most appropriate initial laboratory tests are complete blood count, urinalysis, serum electrolytes, blood urea nitrogen and creatinine concentration, and uric acid.

The most common cardiac cause of hypertension is coarctation of the aorta, but this usually presents in infancy. In older patients with good collateral flow, a "machinery" systolic murmur is typically evident on examination, but in younger patients, the murmur is usually short, systolic, and best heard along the lower sternal border at the third and fourth intercostal spaces. Also, the lower extremities blood pressure will be lower than the upper extremities. Femoral pulses would be weak or absent. Other causes of secondary hypertension in children include drugs (especially corticosteroids, oral contraceptives in sexually active adolescents, and sympathomimetics), lead poisoning, neuroblastoma, and seizure.

(Choice 1) is not indicated for the initial evaluation of a hypertensive child. The purpose of doing this test is to have an accurate measurement of the creatinine clearance, but collection of urine for 24 hours can be very inconvenient to the family. It is also not cost-effective as an initial test.

(Choice 3) Plasma and urine catecholamine levels are used to assess the presence of a catecholamine producing tumor, such as a pheochromocytoma. Pheochromocytoma usually presents with episodic symptoms of palpitation, headaches, flushing, nausea and vomiting, and urinary frequency. Pheochromocytoma is not suspected in this case; therefore, catecholamine levels are not indicated here.

(Choice 4) If elevated, represents either primary or secondary hyperaldosteronism. Clinical manifestations include hypertension, muscle weakness, polydipsia, and polyuria. Laboratory findings include hypokalemia, hyponatremia, and decreased chloride. Therefore, serum electrolytes would be an appropriate initial test for this condition. Serum aldosterone level is not an appropriate initial test in this patient, although it would be indicated later if the patient had the relevant symptoms and characteristic laboratory findings.

(Choice 5) would be used to assess Cushing syndrome, which is another cause of secondary hypertension in children. In children older than

7 years, the most common cause of Cushing syndrome is bilateral adrenal hyperplasia secondary to a pituitary adenoma or ectopic production of corticotropin (ACTH). Children with Cushing syndrome have characteristic moon faces, double chin, buffalo hump, obesity, masculinization, acne, impaired growth, and hypertrichosis.

53. Question

1 points

Category: Pediatrics

A 4-year-old boy from India presents with weakness. His parents note that he has been looking increasingly pale. Hemoglobin electrophoresis demonstrates an abnormal hemoglobin species. Genetic analysis indicates that the patient has the substitution of a valine for a glutamine in the sixth position of the β hemoglobin chain. Which of the following will most likely be seen on his blood smear?

1. ☒ Hypochromic, sickled red blood cells ✓
2. ☐ Hypochromic, spherical red blood cells
3. ☐ Macrocytic, hypochromic red blood cells
4. ☐ Normocytic, hypochromic red blood cells
5. ☐ Normocytic, normochromic red blood cells

INCORRECT ✗

The correct answer is 1.

Sickle cell anemia is very common in the Mediterranean, Africa, and the Indian subcontinent. The valine to glutamine substitution in hemoglobin is the most common genetic defect underlying this illness. This amino acid substitution leads to polymerization of hemoglobin in low oxygen states, causing sickling, and hemolysis in small vessels. The increased use of iron leads to anemia, and the peripheral blood smear shows characteristic sickling of red cells. The cells will also be hypochromic secondary to the low iron content.

(Choice 2) Hereditary spherocytosis is caused by a genetic defect in the cell wall of the erythrocytes. This leads to spontaneous loss of cell wall material, which causes the red cell to assume a spherical shape. Splenic sequestration and destruction follow.

(Choice 3) can be seen in megaloblastic anemia caused by folate and B₁₂ deficiency.

(Choice 4) Most forms of anemia will lead to some decrease in the iron content of the red cells. Thus, normochromic cells would not likely be seen in the smear.

(Choice 5) This patient has sickle cell anemia, and, depending on the severity of his illness, some abnormality should be seen on the smear.

54. Question

1 points

Category: Pediatrics

A 17-year-old girl presents with a 4-week history of intermittent fever, increasing fatigue, generalized myalgia, and swelling of both her knees and ankles. There is a fine erythematous rash on her back, and she has swollen knees and ankles; the remainder of her physical examination is unremarkable. Initial laboratory evaluation shows:

Leukocytes: 11,400cells/mm³

Hemoglobin: 8.8 g/dL

Blood urea nitrogen: 4 mg/dL

Creatinine: 1.4 mg/dL

Glucose: 98 mg/dL

C3 complement: 36 mg/dL (normal >80 mg/dL)

Antinuclear antibody titer: 1:3200

Anti-double-stranded DNA titer: 1:640

Antineutrophil cytoplasmic antibodies: Negative

Urinalysis: Moderate hematuria (50 RBC/hpf)

Moderate proteinuria: (400 mg/dL)

Which of the following is the most likely diagnosis?

1. ☐ Giant cell arteritis
2. ☐ Henoch-Schonlein purpura
3. ☐ Polyarteritis nodosa
4. ☒ Systemic lupus erythematosus ✓
5. ☐ Wegener granulomatosis

INCORRECT ✗

The correct answer is 4.

Systemic lupus erythematosus (SLE) is a multisystemic rheumatologic disease. Its major clinical manifestations include fever, malaise, arthritis, arthralgia, and rash. Anorexia and weight loss are also common. The malar butterfly rash on the face that extends over the bridge of the nose, but spares the nasolabial fold, is very characteristic but is not present in all patients with SLE. Raynaud phenomenon, polyserositis, hepatosplenomegaly, lymphadenopathy, and renal disease also are frequently associated with SLE. Antinuclear antibodies are found in 97% of patients with SLE, but this finding lacks specificity. Antibodies to double-stranded DNA are more specific, and are elevated in active disease. Complement C3 and C4 levels are decreased in patients with active SLE. The American Rheumatism Association has published the criteria for the diagnosis of SLE. At least 4 of the 11 criteria must be met to diagnose SLE.

These criteria can be remembered by a mnemonic SOAP BRAIN MD (see below).

- Serositis
- Oral ulcers
- Arthritis
- Photosensitivity
- Blood (hemolytic anemia, leukopenia, lymphopenia, or thrombocytopenia.)
- Renal (proteinuria or cellular cast)
- ANA
- Immunologic (positive LE preparation, antibody to double-stranded DNA, antibody to Sm antigen)
- Neurologic (seizure or psychosis)
- Malar rash
- Discoid lupus

(Choice 1) also known as Takayasu arteritis, primarily affects the aorta and other large arteries. Symptoms include fever, anorexia, weight loss, and arthritis. Laboratory findings include increased sedimentation rate, weakly positive antinuclear antibody titer, and anti-aorta antibodies.

(Choice 2) is a vasculitis of unknown etiology that occurs most commonly in boys aged between 2 and 8 years. It is characterized by nonthrombocytopenic, palpable purpuras, most commonly in shins, lower extremities, and buttocks. Syndrome also includes arthritis, abdominal pain, and nephritis. The prognosis is good in the absence of significant renal complications.

(Choice 3) is a vasculitis that affects small and medium-sized arteries. It can present with diverse symptoms such as fever, malaise, arthritis, abdominal pain, rash, kidney disorder, ischemic heart pain, testicular pain, peripheral nervous system disorders, lethargy, and weight loss. Antineutrophil cytoplasmic antibodies with peripheral staining (p-ANCA) are positive. Complement levels are normal.

(Choice 5) is characterized by necrotizing granulomatous lesions of the entire respiratory system (airways and lungs). It is also associated with glomerulonephritis. The triad of sinusitis, pulmonary involvement, and glomerulonephritis suggests the diagnosis.

55. Question

1 points

Category: Pediatrics

A 3-month-old infant with a history of chronic constipation presents with fulminant, watery diarrhea over 2 days. The infant is brought to the emergency department with signs of severe dehydration and a distended abdomen. His history is significant for delayed passage of meconium after birth and failure to thrive. Plain x-ray films of the abdomen reveal a massively dilated transverse colon, with the presence of gas in the distal colon. Which of the following is the most likely diagnosis?

1. ☐ Congenital hypothyroidism
2. ☐ Meconium ileus
3. ☐ Necrotizing enterocolitis
4. ☒ Toxic enterocolitis
5. ☐ Volvulus

INCORRECT ❌

The correct answer is 4.

The most likely cause of this infant's presentation is toxic enterocolitis (toxic megacolon), which is most often seen in the setting of Hirschsprung disease. The disease is suggested in this infant by the delayed passage of meconium, chronic constipation, and the failure to thrive. When unrecognized, putrefaction and development of colitis can occur. In its severest form, it presents as toxic megacolon. The resultant diarrhea, with potentially fatal fluid loss, is due to bacterial overgrowth in the dilated bowel with resulting production of bacterial toxins. Although fluid replacement and antibiotics are important in the management of these cases, the most helpful therapeutic intervention is prompt surgical resection of the involved bowel with formation of a colostomy.

(Choice 1) Infants with congenital hypothyroidism have a history of constipation. However, other features of hypothyroidism are lacking in this case. Classic features include prolonged jaundice, umbilical hernia, macroglossia, hypotonia, and failure of the posterior fontanelle to close.

(Choice 2) occurs in infants with cystic fibrosis who fail to pass meconium in the first few days of life.

(Choice 3) is a feared complication of the newborn period, with abdominal distension and sepsis.

(Choice 5) Volvulus neonatorum is usually due to a defect in the normal rotation of the bowel in which the caecum remains high, often with a congenital band passing across the duodenum. This results in an intestinal obstruction and vessel occlusion at the base of the mesentery. Clinically, the features of intestinal obstruction predominate, with vomiting, abdominal pain, and constipation. Abdominal radiograph shows a "comma sign" or "beak sign" with absence of distal bowel gas.

56. Question

Category: Pediatrics

1 points

A 7-year-old Caucasian male with a history of peanut allergy develops sudden shortness of breath after lunch at school. On physical examination, his blood pressure is 70/50 mmHg and his heart rate is 130/min. Inspirations and expirations are labored and noisy. Which of the following is the best initial treatment?

1. ☐ Albuterol nebulizer
2. ☒ Subcutaneous epinephrine injection ✓
3. ☐ Intravenous steroids
4. ☐ Intravenous diphenhydramine
5. ☐ Endotracheal intubation

INCORRECT ✗

The correct answer is 2.

This patient has developed labored breathing, tachycardia and hypotension. Given his history of peanut allergy, these findings are most likely an anaphylactic reaction to peanut proteins. His stridor and wheezing are most likely due to mucosal edema and bronchospasm, respectively. Food allergies are the major cause of pediatric outpatient anaphylaxis, and peanut allergies account for the majority of fatal and near fatal reactions. In anaphylaxis, there is an IgE-mediated release of histamine and leukotrienes from mast cells and basophils, which cause relaxation of vascular smooth muscle and contraction of bronchial and intestinal smooth muscle. Epinephrine is the first-line treatment for acute anaphylaxis when ventilation is adequate. Epinephrine is an α_1 agonist that vasoconstricts to raise blood pressure. It also works as a β_2 -agonist, causing bronchial smooth muscle relaxation to counter bronchospasm.

(Choice 1) Albuterol, a $\beta_2 > \beta_1$ agonist, can adjunctively help relieve bronchoconstriction but does not affect the other systemic symptoms of anaphylaxis, like hypotension.

(Choice 3) Corticosteroids can be of benefit in cases of airway constriction caused by late-stage inflammation, but do not have the sort of immediate effect required in this emergency situation.

(Choice 4) Antihistamines – both H_1 and H_2 blockers – can benefit common allergy sufferers, but do not reverse the hypotension and bronchospasm of anaphylaxis and therefore are an inappropriate first-line choice here.

(Choice 5) In an anaphylactic patient with respiratory failure, intubation should be the first step. Signs of respiratory failure like cyanosis and obtundation are not yet present in this patient.

57. Question

1 points

Category: Pediatrics

A 2-week-old infant is brought to the emergency department because of diarrhea, poor feeding, abdominal distention, and vomiting for the past 3 days. Physical examination is pertinent for temperature (39°C [102.2°F]), cataracts, hepatomegaly, and jaundice. The patient has poor skin turgor and a capillary refill of 3 seconds. The physician suspects that the infant is septic and has galactosemia. Which of the following would be the most probable pathogen involved in the sepsis?

- ☐ Group B streptococcus
- ☐ *Listeria monocytogenes*
- ☒ *Escherichia coli* ✓
- ☐ *Staphylococcus aureus*
- ☐ *Pneumococcus* (*Streptococcus pneumoniae*)

INCORRECT ✗

The correct answer is 3.

A strong association exists between galactosemia and *Escherichia coli* sepsis, which may be a presenting feature in infants with galactosemia. Classic galactosemia results from a deficiency of galactose-1-phosphate uridylyltransferase. Galactose, derived from lactose (milk sugar), is not metabolized past galactose-1-phosphate, which then accumulates and damages the liver, kidney, and brain. Galactitol, a polyol byproduct of galactose, also can accumulate, causing cataracts. Among other manifestations are jaundice, hepatomegaly, hypoglycemia, vomiting, aminoaciduria, and failure to thrive. Early diagnosis is important, and dietary management contributes to a good prognosis.

(Choices 1,2,4 & 5) Although group B streptococcus, *Listeria monocytogenes*, *Staphylococcus aureus*, and pneumococcus (*Streptococcus pneumoniae*) are also pathogens seen in neonatal sepsis, only *E. coli* has a strong association with galactosemia.

58. Question

1 points

Category: Pediatrics

A 11-month-old boy is brought to the emergency department by his parents. The child has a fracture of the right femur. The father reports this was sustained as a result of falling out of the crib. The child is also noted to have bruises on his shoulders and back. The rest of his examination is unremarkable. Which of the following is the most appropriate next step in diagnosis?

1. ☐ Social services consult
2. ☐ Chest x-ray
3. ☐ CT of the head
4. ☒ Funduscopic exam ✓
5. ☐ Lumbar puncture

INCORRECT ✗

The correct answer is 4.

This child is presenting with multiple atypical injuries, and child abuse must be suspected. Injuries from being shaken and subtle head trauma are common findings in abused children. Retinal hemorrhage may be noted. Funduscopic examination is imperative before other measures are undertaken.

(Choice 1) A social services consult will probably be needed if there is suspicion of child neglect and abuse. In such situations, the child may be placed in a foster home.

(Choice 2) may be needed to assess for rib fractures. Although this ultimately may be required to complete the child's evaluation, it is not needed initially. Nondisplaced fractures should heal without intervention.

(Choice 3) may be needed to assess for brain injury, including intracranial hemorrhage. Cranial fractures may be seen, and coup/contrecoup contusions may be noted as well.

(Choice 5) may be needed if there is strong suspicion of head trauma and the child is exhibiting neurologic deficits. Even if the head CT is negative, the lumbar puncture may indicate the presence of a hemorrhage. This should be done with caution, since the intracranial pressure may be elevated and there is risk of herniation.

59. Question

1 points

Category: Pediatrics

A 1-year-old child is brought in for a regular "well baby" check-up. The child appears to have strabismus. The reflection of a bright light from the ceiling of the examination room comes from a different place in each eye. The family explains that the child has always looked that way, and there has been no recent change in the appearance of his eyes. Which of the following is the most effective management?

1. ☐ No treatment unless the condition has not resolved spontaneously by age 7
2. ☐ Corrective lenses
3. ☐ Each eye patched for a month at a time, alternating sides
4. ☐ Surgical correction whenever he is old enough to decide whether he wants it for cosmetic reasons
5. ☒ Surgical correction as soon as it is practical to do it ✓

INCORRECT ✗

The correct answer is 5.

This is not an emergency, but strabismus must be corrected as soon as possible to enable the brain to learn to process images from both eyes. Otherwise, images from one side are suppressed, by age 7, permanent cortical blindness will occur in the suppressed side (amblyopia).

(Choice 1) Spontaneous correction does not occur. It is an often-quoted myth, probably stemming from the fact that some children with a broad base of the nose appear to be cross-eyed and then "grow out of it." They never had strabismus, and, if properly examined, the corneal reflection of a bright light would be seen coming from the same place in each eye.

(Choice 2) Corrective lenses are the treatment for a different type of strabismus: if a child has had normal alignment until he reaches an older age, and then gets "cross-eyed" whenever he focuses on a nearby object. This type of patient has an accommodation problem that can be corrected with appropriate lenses.

(Choice 3) Eye patches are used to force the brain to process "images from the "bad eye" while preparations are made for surgical correction. Only the "good eye" is patched, and the notion of alternating patches would not be effective and could not be the only treatment.

(Choice 4) Delayed correction for cosmetic purposes only would be appropriate if the strabismus had not been diagnosed or treated during infancy, so that when the patient was first seen (after the age of 7), he already had irreversible cortical blindness.

60. Question

1 points

Category: Pediatrics

A 10-year-old girl comes to medical attention because of recurrent attacks of wheezing and dyspnea. The attacks occur mostly at home or, if outdoors, soon after exercise. Exacerbations are noted in springtime. The severity of symptoms is mild. Pulmonary function tests show that peak expiratory flow (PEF) and forced respiratory volume per second (FEV1) are reduced during an attack but are relatively normal during symptom-free intervals. Height and weight are in the 60th percentile. Complete blood count shows 8% eosinophils; all other parameters are normal.

Cutaneous testing shows the patient to be allergic to a variety of allergens, including dust mites, animal dander, and several pollens. Which of the following is the most effective step in management?

1. ☐ Avoidance of exercise
2. ☒ Avoidance of respiratory irritants, such as cigarette smoke ✓
3. ☐ Use of a humidifier at home
4. ☐ Use of air cleaners at home
5. ☐ Administration of multiple-drug regimens

INCORRECT ✗

The correct answer is 2.

The most crucial step in the management of asthma is avoidance of triggering factors, e.g., allergens. Unfortunately, it is difficult to avoid specific types of allergens, such as pollens. Specific measures to eliminate or reduce exposure to dust mites and animal dander at home lead to reduced frequency of attacks and hospitalization rates. Regardless of the allergens involved, elimination of respiratory irritants, especially cigarette smoke, is of crucial importance. The bronchial tree of asthmatic patients is highly reactive to any form of chemical or physical irritation. Thus, exposure to passive smoke should be absolutely avoided.

(Choice 1) is not an appropriate measure, although exercise frequently triggers asthmatic attacks. The efficacy of anti-asthma measures can be evaluated by observing how the child can sustain adequate forms of exercise.

(Choice 3) Use of a humidifier at home will favor growth of dust mites and consequently increase the concentration of these allergens in the environment.

(Choice 4) has not been shown to be uniformly effective in getting rid of dust mites.

(Choice 5) is not a recommended strategy in the pharmacologic treatment of asthma. In fact, the fewest number of drugs at the lowest effective doses should be used. Typically, a regimen of one drug (a bronchodilator or a inhaled corticosteroid) for mild-moderate cases or two drugs for more severe cases is sufficient to control asthma exacerbations.

61. Question

1 points

Category: Pediatrics

An infant is brought to the office for health maintenance visit. On examination, the infant turns when her name is called. She is able to say "mama." Her mother mentions that she also says "dada" at home. She is able to look for her mother when she gets frightened. She also waves bye-bye to the doctor when the doctor steps out of the examination room. What age of this child is most consistent with these developmental milestones?

1. ☐ 3 months
2. ☐ 5 months
3. ☐ 7 months
4. ☒ 9 months
5. ☐ 11 months

INCORRECT ❌

The correct answer is 4.

Developmental assessment is essential in pediatrics health maintenance. It should be included in every health supervision visit. A developmental screening includes assessment of the following four areas: neurodevelopment, cognitive development, language development, and psychosocial development. The ability to make repetitive consonant sounds, such as "mama" or "dada," is learned by 8-9 months. At that time, the infant can also recognize his or her own name. In term of social cognition, an infant will seek reassurance from a familiar person, usually parents or caretakers, when frightened. By 9 months, an infant can also wave bye-bye.

(Choice 1) Neurodevelopment is more important. At this time, an infant can raise his or her head when placed on a firm surface. The infant may be able to laugh at pleasurable social contacts.

(Choice 2) An infant can often be pulled from a sitting to a standing position. The infant can also support his or her weight on extended legs.

(Choice 3) The infant can make repetitive vowel sounds but is not able to recognize his or her own name yet. A prone infant at this age is able to pivot in pursuit of an object.

(Choice 5) The infant enjoys playing ball with the examiner. He or she can also use a few words other than "mama" or "dada."

62. Question

1 points

Category: Pediatrics

A female neonate is undergoing an examination after birth. She was born to a 33-year-old primigravid mother at term via a normal spontaneous vaginal delivery. The pregnancy was uncomplicated, except for a positive maternal group B Streptococcus culture at 36 weeks' gestation, for which the mother received penicillin during labor. The infant's APGAR scores are 8 at 1 minute and 9 at 5 minutes. The mother notices that the infant has prominent labia and a dull pink vaginal epithelium. Which of the following is the most likely cause of the appearance of the infant's genitalia?

1. ☒ Exposure to maternal estrogen ✓
2. ☐ Exposure to penicillin
3. ☐ Infection with Chlamydia
4. ☐ Infection with group B Streptococcus
5. ☐ Sexual abuse

INCORRECT ✗

The correct answer is 1.

At birth, a neonate will typically exhibit some signs of exposure to maternal estrogen. Two of these signs include prominent labia and a dull pink vaginal epithelium. As estrogen levels fall, the labia will become less prominent and the vagina will take on a brighter red appearance. In the absence of other signs or symptoms of disease, these findings are normal and do not require further evaluation.

(Choice 2) will almost never cause findings isolated to the labia and vagina.

(Choice 3) can lead to neonatal pneumonia and eye infection.

(Choice 4) can cause respiratory distress, pneumonia, meningitis, and sepsis in the newborn. It does not typically cause findings isolated to the genitalia.

(Choice 5) almost always must be considered whenever there are complaints or findings regarding an infant's genitalia. In this case, however, the infant is newborn-only minutes old. Therefore, sexual abuse would not be in the differential diagnosis.

63. Question

1 points

Category: Pediatrics

A 12-year-old boy comes to the emergency department with severe shortness of breath, cough, and fever. He was diagnosed with cystic fibrosis when he was five years old. He is currently not taking any medications. His temperature is 39.4 °C (103 °F), blood pressure is 80/40 mm Hg, pulse is 120/min, and respirations are 30/min. He is excessively using his accessory respiratory muscles. Lung auscultation reveals bilateral lower lobe crackles and expiratory wheezing. Pulse oximetry shows SaO₂ of 85% that improves to 92% after oxygen supplementation. Which of the following antibiotics or combinations is the best choice for this patient?

1. ☒ Intravenous ceftazidime and gentamicin ✓
2. ☐ Intravenous vancomycin and gentamicin
3. ☐ Intravenous imipenem/cilastatin
4. ☐ Intravenous clindamycin
5. ☐ Oral ciprofloxacin

INCORRECT ✗

The correct answer is 1.

This cystic fibrosis patient is suffering from an acute exacerbation of lung disease, and his presentation (i.e., prominent shortness of breath, excessive use of accessory respiratory muscles, hypotension J is suggestive of a severe attack. The most common etiologic agent is *Pseudomonas aeruginosa*, although a variety of other agents have been implicated (e.g., *S. aureus* and *H. influenzae*). The treatment is rapid administration of empiric antibiotics, and the usual choice is a combination of two agents with coverage against *Pseudomonas aeruginosa*, (i.e., ceftazidime or a penicillin derivative such as ticarcillin +an aminoglycoside such as amikacin or gentamicin J.

(Choice 3) The other drugs that are effective against *Pseudomonas* are ceftipime and imipenem/cilastatin combination.

Imipenem/cilastatin is effective against *Pseudomonas aeruginosa*, but is used as a second-line drug. Double coverage (combination antibiotics J is important, because of the remarkable resistance to this organism to most antibiotics, and its ability to develop further resistance during the course of therapy.

(Choice 2) The addition of an anti-staphylococcal antibiotic (e.g., nafcillin or vancomycin) depends on the results of the culture.

(Choice 5) Oral ciprofloxacin covers *Pseudomonas aeruginosa*, but it is not appropriate for severe exacerbations, wherein intravenous antibiotics are usually employed. The new generation fluoroquinolones (e.g., levofloxacin, gatifloxacin) do not have good coverage against *Pseudomonas*.

(Choice 4) Clindamycin is effective against anaerobes, and has little role in the treatment of such patients.

64. Question

1 points

Category: Pediatrics

A 4-day old preterm male neonate is being managed in the neonatal intensive care unit. He was born in the 34th week of gestation via lower segment caesarian section. On the first day of life, he developed tachypnea, grunting and nasal flaring. Chest x-rays showed a ground glass appearance of both lungs. He underwent endotracheal intubation and therapy with intravenous fluids, positive pressure ventilation, and intravenous antibiotics. He consequently showed remarkable improvement; however, he is currently beginning to have increased gastric residues. What is the most likely cause for this child's increased gastric residues?

1. ☐ Trachea-esophageal fistula
2. ☒ Necrotizing enterocolitis ✓
3. ☐ Duodenal atresia
4. ☐ Meconium aspiration syndrome
5. ☐ Pyloric stenosis

INCORRECT ✗

The correct answer is 2.

Increased gastric residues in a preterm neonate is highly suspicious for necrotizing enterocolitis. The condition usually presents at 3-10 days after birth, and is due to bowel wall injury resulting from perinatal asphyxia. The diagnosis requires a high index of suspicion.

Treatment depends on the degree of bowel involvement and severity of presentation. Preterm neonates are at high risk for other complications such as hyaline membrane disease, which presents with respiratory distress and ground glass appearance of the lungs on chest x-rays.

(Choice 1) Trachea-esophageal fistula presents with excessive drooling, choking, coughing, and cyanosis due to aspiration of feedings.

(Choice 3) Bilious vomiting without abdominal distention is the cardinal sign for duodenal atresia.

(Choice 4) Clinical features of meconium aspiration syndrome include thick, greenish amniotic fluid in the mouth and throat at birth, accompanied by cyanosis and respiratory distress.

(Choice 5) Pyloric stenosis is characterized by projectile nonbilious vomiting and palpation of an olive-shaped mass immediately after the infant has vomited.

65. Question

1 points

Category: Pediatrics

10-day course of penicillin G. His temperature is 38.8 °C(102 °F) and pulse rate is 102/min. On examination, his right knee joint is swollen and erythematous, with a decreased range of motion. The laboratory findings are significant for leukocytosis and an elevated ESR. X-rays of both the right hip and knee joints are normal. What is the most appropriate treatment for the patient's condition?

1. ☐ Bed rest with the right knee joint in a position of comfort
2. ☐ Repeat a ten-day course of penicillin G
3. ☐ Bed rest & a ten-day course of penicillin G
4. ☐ Bed rest & aspirin
5. ☒ Right knee arthrocentesis & intravenous nafcillin ✓

INCORRECT ✗

The correct answer is 5.

This child has features suggestive of septic arthritis. Septic arthritis can occur following skin or upper respiratory tract infections.

Arthrocentesis is both diagnostic and therapeutic, and is mandatory in all suspected cases. Blood cultures and synovial fluid cultures must be obtained prior to administering the antibiotics. Empiric antibiotic therapy (IV nafcillin or IV cefazolin) should be started with pending synovial fluid cultures.

(Choice 1) Bed rest for 7 days with the knee joint in a position of comfort is the treatment for transient synovitis, not septic arthritis.

(Choices 2 & 3) The patient has received an adequate course of penicillin for pharyngitis; therefore, repeating the course is not required. Moreover, penicillin G is not the drug of choice for the empiric treatment of septic arthritis. The most common cause of septic arthritis in a native joint is *Staphylococcus aureus*. Almost 99% of the *staphylococcus* strains are resistant to penicillin. The commonly used empirical antibiotics are nafcillin or cefazolin. Vancomycin should be used if the suspicion for methicillin resistant *staphylococcus aureus* (MRSA) is high.

(Choice 4) Aspirin is not usually used in children because of the risk of Reye's syndrome.

66. Question

1 points

Category: Pediatrics

A pregnant woman has premature rupture of membranes. Her baby is born 3 days later, at 37 weeks' gestation. The 5-minute APGAR score is 4. Lung sounds are reduced, and the infant appears to be in respiratory distress. Peripheral blood smear with differential counts demonstrates a neutrophil count of 30,000/mL, with toxic granules evident in many neutrophils. Gram stain of huffy coat demonstrates small gram-positive cocci in chains. Which of the following is the most likely causative organism?

1. ☐ Group A Streptococcus
2. ☒ Group B Streptococcus ✓
3. ☐ Methicillin-resistant Staphylococcus aureus
4. ☐ Methicillin-sensitive Staphylococcus aureus
5. ☐ Neisseria meningitidis

INCORRECT ✗

The correct answer is 2.

This is neonatal sepsis, which in the first few days of life is most likely to be due to group B Streptococcus or gram-negative enteric organisms. Physicians should maintain a high index of suspicion, since neonatal sepsis may be subtle or nonspecific in its symptoms ("not doing well," respiratory distress, apnea, bradycardia, seizures, jaundice). Gram stain of the buffy coat from a blood sample may be particularly helpful in establishing the diagnosis. Predisposing conditions include obstetric complications, toxemia, and maternal infection. (Choice 1) does not commonly infect infants; it causes sore throats, pneumonia, and meningitis in older children.

(Choices 3 & 4) Staphylococcus aureus, in either its methicillin-sensitive or methicillin-resistant forms, can cause skin pustules, sepsis, pneumonia, and meningitis in infants, but would be described as gram-positive cocci in clusters rather than chains.

(Choice 5) is a gram-negative diplococcus that can cause meningitis and respiratory infections in children but is not common in neonates.

67. Question

1 points

Category: Pediatrics

A 2-year-old boy is brought to the clinic by his mother because he is lethargic and not as active as the other kids on their block. He has difficulty walking and getting up, and he always appears tired. His mother thinks that she has a relative with a disorder that causes weakness. On examination, the child is alert but quiet. His calves appear hypertrophied. His reflexes are depressed. When he gets up from the floor, he uses his hands to climb on his legs to assume an upright position. What is the most likely diagnosis of this child?

1. ☐ Myasthenia gravis
2. ☐ Dermatomyositis
3. ☒ Duchenne muscular dystrophy ✓
4. ☐ Polio myelitis
5. ☐ Fragile X syndrome

INCORRECT ✗

The correct answer is 3.

This boy presents with proximal muscle weakness, pseudo hypertrophied calf muscles, decreased reflexes, and a positive Gower's sign (uses hands to walk up the legs and assume an upright position). His features are very characteristic of Duchenne muscular dystrophy. Other features include obesity, mental retardation and cardiomyopathy. Dystrophin is usually absent. EMG shows myopathic pattern. Serum CK levels are very high (4000-5000 IU). The diagnosis is confirmed by muscle biopsy and family pedigree analysis. The current treatment is supportive, although gene therapy trials are underway. The prognosis is poor, with most patients wheelchair-bound by 12 years of age.

(Choice 1) Myasthenia gravis symptoms may fluctuate, and may be purely ocular or generalized. Symptoms may include diplopia, ptosis, and muscle fatigue with normal deep tendon reflexes. Ten to fifteen percent of patients have a thymoma; hence, a CT scan is done to check for a thymoma. The diagnosis is confirmed by a positive edrophonium test (temporary improvement after the administration of edrophonium).

(Choice 2) Dermatomyositis has a gradual onset in adult life, and is more common in females. Skin changes may precede or follow muscle findings. There may be a rash on the eyelids, nose, cheeks, trunk and extremities. There may be an associated vasculitis in 25% of cases. Muscle biopsy is diagnostic.

(Choice 4) Polio may be a subclinical or a mild abortive illness. The individual may have a non-specific febrile illness. Some individuals may have flaccid paralysis, absent deep tendon reflexes and normal sensation. There is no hypertrophy of the muscles. Approximately 10-25% may recover for up to 6 months.

(Choice 5) Fragile X syndrome is an X-linked recessive disorder and is a major cause of mental retardation in males. The typical features include facial overgrowth and Macroorchidism. Patients may also be hyperactive or autistic. Patients do not have any muscular deficits.

68. Question

1 points

Category: Pediatrics

A child is at his pediatrician's office for a well-child check. The patient has been well according to the parents, and there are no complaints. On physical examination, no abnormalities are noted. Additionally, the pediatrician performs a developmental screen on the patient. He finds that the child is able to say his first and last name and play interactive games. The patient's mother reports that the patient is toilet trained and is able to pedal a tricycle. He is capable of copying a cross and a circle, but cannot copy a square or triangle. He is also not able to catch a bounced ball or dress without supervision. Which of the following is the most likely age of this child?

1. ☐ 1 year old
2. ☐ 2 years old
3. ☒ 3 years old ✓
4. ☐ 4 years old
5. ☐ 5 years old

INCORRECT ✗

The correct answer is 3.

Most 3-year-old children are toilet-trained, play interactive games, give their first and last names, and pedal a tricycle. At age 3, most children should be able to copy a circle, and some will be able to copy a cross.

(Choice 1) A 1-year-old child would not be able to pedal a tricycle, give his first and last names, or copy a circle or a cross. At age 1, a child should be able to stand momentarily, say "dada" and "mama," and bang two cubes held in his hands.

(Choice 2) Although some 2-year-old children may be able to accomplish the tasks noted in the scenario, they would not be able to hop on one foot.

(Choice 4) Most 4-year-old children are able to dress without supervision, hop on one foot, and catch a bounced ball.

(Choice 5) A 5-year-old child would be able to copy a triangle.

69. Question

Category: Pediatrics

1 points

A 12-year-old boy is brought to the physician because of pain in his right leg for the past 3 weeks. The pain frequently occurs at night and is localized to the tibia, a few centimeters below the knee. The mother reports that the pain is promptly relieved by aspirin and that the child has had no fever. Examination reveals no tissue swelling or redness about the site of pain. X-ray films show a 1-cm radiolucent focus in the tibial cortex surrounded by marked bone sclerosis. Which of the following is the most likely diagnosis?

1. ☐ Aneurysmal bone cyst
2. ☐ Enchondroma
3. ☐ Ewing sarcoma
4. ☒ Osteoid osteoma ✓
5. ☐ Osteosarcoma

INCORRECT ✗

The correct answer is 4.

Nocturnal bouts of bone pain in a child that are promptly relieved by aspirin should raise the suspicion of osteoid osteoma. The diagnosis is supported by the radiographic finding of a radiolucent nidus surrounded by a wide rim of osteosclerosis in a typical location. Typically, this benign tumor is located in the metaphyseal cortex of long bones, especially the femur and tibia, and bones of the hands and feet. Osteoid osteoma consists of haphazardly arranged bone trabeculae separated by fibrovascular tissue. It is treated with surgical resection.

(Choice 1) most commonly affects the metaphysis of the tibia and femur close to the knee in children and young adults. It is a spongy hemorthagic multilocular cyst that expands the bone, eroding the cortex. X-ray films show an eccentric area of osteolysis, which is well demarcated and surrounded by thinned cortex. It manifests with pain and functional impairment of the adjacent joint.

(Choice 2) Enchondroma, or chondroma, is a benign cartilaginous tumor that most frequently develops in the medullary cavity of phalangeal, metacarpal, and metatarsal bones. X-ray films show an osteolytic area in the diaphysis surrounded by thinned cortex. Frequently asymptomatic, it may manifest with pain.

(Choice 3) is a malignant tumor occurring most frequently in long bones, especially the femur and humerus. Radiographically, the tumor gives rise to a moth-eaten area of osteolysis associated with a periosteal reaction and an onion-skinning pattern. It manifests with pain and systemic symptoms, e.g., fever, leukocytosis, and elevated erythrocyte sedimentation rate.

(Choice 5) is the most common primary malignant tumor of the bone. It most frequently affects males between 10 and 20 years of age. Preferential locations include the metaphysis of the tibia and femur around the knee and the upper metaphysis of the humerus. Intense local pain, swelling, and pathologic fractures are clinical manifestations. X-ray films show destruction of the cortical bone with a periosteal reaction manifesting with the Codman triangle.

70. Question

1 points

Category: Pediatrics

A 2-year-old male child from a Vietnamese immigrant family is brought to the emergency department by a Child Protective Services (CPS) worker for suspected physical abuse. The worker states that, earlier that morning, she received a call from a day care facility that the child attends. According to the CPS worker, the day care employee stated that she thought that the child had been physically abused. The physician performs a thorough history and physical examination to determine evidence of maltreatment. Which one of the following would be specific evidence of child abuse in this case?

1. ☐ Bruises on the back in a fir-tree pattern from "coining"
2. ☐ A blue, nontender macular lesion in the presacral area
3. ☐ Weight less than the fifth percentile
4. ☐ Bruised knees
5. ☒ A circular burn restricted to the buttocks ✓

INCORRECT ✗

The correct answer is 5.

Specific evidence of child abuse includes old healed fractures, belt marks, bruises on the buttocks or lower back (areas where children are unlikely to harm themselves), subdural hematomas, and rupture of internal organs. Hot-water-dunking burns occur when a parent holds the child's thigh against the abdomen and places the buttocks and perineum in scalding water. This causes a circular type of burn restricted to the buttocks. The hands and feet are spared, which is not compatible with falling into a tub. Evidence of sexual abuse includes genital or anal trauma, sexually transmitted disease, and urinary tract infection.

(Choice 1) Coin rubbing is a non-abusive healing practice of the Vietnamese that involves vigorous stroking of the skin of a febrile child, producing a peculiar bruising pattern.

(Choice 2) A blue, nontender lesion in the presacral area is a "Mongolian spot" often seen in African American, Oriental, and East Indian infants. Unlike a bruise, the Mongolian spot does not change color.

(Choice 3) Although caloric insufficiency from child abuse may explain weight below the fifth percentile for age, it is not the sole reason for failure to thrive.

(Choice 4) are commonly seen in children of this age because of falls and are not specific evidence of child abuse.

71. Question

1 points

Category: Pediatrics

A 4-day-old, 1,500 g (3lb 5oz) premature infant recovering from a respiratory distress syndrome is noted to have bounding peripheral pulsations and a hyperactive precordium. A continuous machinery murmur is most audible at the left infraclavicular area. Left ventricle hypertrophy (LVH) is present on the electrocardiogram (ECG). The chest x-ray film shows slight enlargement of the heart and increased pulmonary venous markings. Which of the following is the most likely diagnosis?

1. ☒ Patent ductus arteriosus ✓
2. ☐ Atrial septal defect (ASD)
3. ☐ Ventricular septal defect (VSD)
4. ☐ Pulmonic stenosis
5. ☐ Tetralogy of Fallot

INCORRECT ✗

The correct answer is 1.

A patient with patent ductus arteriosus has a continuous machinery murmur. Bounding pulses with hyperactive precordium may be found on physical examination. Although the electrocardiogram (ECG) is usually not helpful, it may show left ventricular hypertrophy (LVH).

(Choice 2) Patients with atrial septal defect (ASD) have a widely split and fixed second heart sound.

(Choice 3) Patients with ventricular septal defect (VSD) have a harsh holosystolic murmur heard best over the lower left sternal border.

(Choice 5) is characterized by the presence of a VSD, right ventricular hypertrophy, pulmonic stenosis, and overriding aorta. The murmur of tetralogy of Fallot is a long, systolic ejection murmur most audible at the mid-left sternal border.

(Choice 4) In patients with pulmonic stenosis, there is a high-pitched systolic ejection murmur, most audible at the upper left sternal border, which transmits fairly well to the back. A systolic thrill may be present at the upper left sternal border and, rarely, in the suprasternal notch.

72. Question

Category: Pediatrics

1 points

A 15-month-old child is brought to the office by his mother because he cries incessantly when he is left at the local day care. His mother is concerned because he is "a happy child at home and loves playing with his other siblings." His growth pattern is normal, and has achieved all the age-appropriate physical and mental milestones. What is the most likely diagnosis?

1. ☐ Separation anxiety disorder
2. ☐ Stranger anxiety
3. ☐ Social anxiety disorder
4. ☒ Autism
5. ☐ Rett's disorder

INCORRECT ❌

The correct answer is 4.

Stranger anxiety is a normal phenomenon that occurs in most infants. It is the normal distress that they experience when exposed to unfamiliar people. It usually starts anywhere between 6-8 months, and peaks at 12-15 months.

(Choice 1) Separation anxiety disorder is the excessive anxiety associated with separation from home or a loved one. Individuals suffering from this disorder experience extreme discomfort on or at the thought of separation from an individual they are attached to. They go to extremes to avoid such situations. This could have been an acceptable diagnosis in an older child, but in this child of 15 months, stranger anxiety is more likely.

(Choice 3) Social anxiety or phobia is a significant fear associated with exposure to one or more social situations. Exposure to the feared situations provokes extreme anxiety, and the individual makes great efforts to avoid them.

(Choice 4) Autism is characterized by qualitative and quantitative disturbances of communication, coupled with performance of certain stereotyped activities. Individuals also tend to engage in solitary activities. These features are not present in the abovementioned patient, who is described as "playful with his siblings".

(Choice 5) Rett's disorder is characterized by a deterioration of previously acquired skills, beginning at the age of five months. The presence of normal mental and physical milestones in the abovementioned patient makes this diagnosis very unlikely.

72. Question

1 points

Category: Pediatrics

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INCORRECT 

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73. Question

1 points

Category: Pediatrics

A pediatrician is called to attend a delivery of a 35-year-old primigravida. According to the patient and review of the medical record, the patient has had good prenatal care. And, except for the development of diabetes during the second trimester, the pregnancy has been uneventful. Cultures for sexually transmitted diseases are documented as negative. After many hours of labor the patient delivers an infant by spontaneous vaginal delivery with Apgar scores of 8 at 1 minute and 9 at 5 minutes. Birth weight is 4.5 kg (10 lb). Which of the following injuries is most likely to be found in this newborn?

1. ☐ Cephalhematoma
2. ☐ Facial nerve paralysis
3. ☐ Intraventricular hemorrhage
4. ☐ Brain contusion
5. ☒ Fractured clavicle ✓

INCORRECT ✗

The correct answer is 5.

The clavicle is the most commonly fractured bone in infants during delivery. This fracture is more common in macrosomic newborns (e.g., infants of diabetic mothers). The newborn generally does not move the arm, giving an absent Moro reflex on the affected side. Crepitus is felt on palpation. Treatment is usually unnecessary, and the prognosis is good.

(Choice 1) is a hemorrhage confined within the area of the affected cranial bone. Cephalhematomas require no treatment, although phototherapy may be necessary to ameliorate hyperbilirubinemia.

(Choice 2) may result from pressure over the facial nerve in utero or from forceps during delivery. A patient with facial nerve palsy will have an eye that cannot be closed, an absent nasolabial fold, a corner of the mouth that droops, and a smooth forehead on the affected side. Prognosis depends on the extent of injury to the nerve.

(Choice 3) is caused by rupture of the germinal blood vessels. Predisposing factors are prematurity, hypoxic injury, respiratory distress syndrome, reperfusion injury, and change in volume of cerebral circulation, compromised vascular integrity, hypervolemia (hypertension), and pneumothorax. The incidence of intraventricular hemorrhage increases with decreasing birth weight.

(Choice 4) may be associated with a skull fracture.

74. Question

1 points

Category: Pediatrics

A 1-year-old boy is brought to the pediatrician for a routine visit. While talking to his mother, she reports that they moved into an old house several months ago and are in the process of renovating. The child eats table food and drinks whole milk. His height, weight, and head circumference are all at the 50th percentile for his age. Physical examination is normal. The results of a fingerstick blood test are shown below:

Hemoglobin: 10.5 g/dL

Hematocrit: 30.0%

Lead level: 12 µg/dL (Normal < 10 µg/dL) Which of the following is the most appropriate next step in management?

- ☐ Remove the child from the house
- ☒ Check a serum lead level ✓
- ☐ Initiate chelation therapy with dimercaprol
- ☐ Initiate chelation therapy with dimercaptosuccinic acid (DMSA, succimer)
- ☐ Recheck lead level in one month

INCORRECT ✗

The correct answer is 2.

Lead poisoning should be screened for routinely by all pediatricians. Children are considered at risk for lead poisoning if they live in an old house, especially if there is peeling paint or during renovation projects. Other risk factors include lead pipes, living near a battery recycling plant, having a parent who works with batteries or pottery, or having a playmate or sibling with a history of lead poisoning. Capillary blood specimens (using a fingerstick) are widely used in the screening for lead poisoning. However, false positive results are common for various reasons. Therefore, further work-up with a confirmatory serum venous blood level is required if a screening capillary lead level is > 10 µg/dL. If the serum lead level is > 10 µg/dL as well, intervention is needed.

(Choice 1) Removing the child from the house would be the most important step in reducing exposure if the diagnosis of lead poisoning is confirmed.

(Choices 3 & 4) Chelation therapy with either dimercaprol or dimercaptosuccinic acid (DMSA, succimer) is typically used when lead levels are extremely elevated (>45 µg/dL).

(Choice 5) Rechecking the lead level in one month is appropriate for children with mildly elevated lead levels A. Remove the child from the house.

75. Question

1 points

Category: Pediatrics

A 7-year-old boy is brought to the physician because of persistent nasal obstruction for 6 months. There is no personal or family history of allergic disorders. Examination of the nasal fossae reveals bilateral ethmoidal polyps that protrude into the middle meatus and nasal cavity. Which of the following is the most appropriate next step in diagnosis?

- ☐ Cutaneous allergen testing
- ☐ Excisional biopsy
- ☐ Nasal provocation testing
- ☒ Pilocarpine iontophoresis sweat test ✓
- ☐ Radioallergosorbent test (RAST)

INCORRECT ✗

The correct answer is 4.

Nasal polyps are not neoplasms, but rather a florid hyperplastic response of nasal and paranasal mucosa to chronic inflammation. Allergic rhinitis/sinusitis is the most common underlying condition. However, presence of polyps in children should raise the possibility of cystic fibrosis. This should be promptly investigated by a pilocarpine iontophoresis sweat test, which would show elevated sweat chloride in patients with cystic fibrosis. Up to 20% of cystic fibrosis patients ultimately develop nasal polyps.

(Choices 1 & 5) Cutaneous allergen testing and detection of allergen-specific serum IgE by in vitro RAST constitute the proper course of action when there is clinical evidence of an allergic nature of nasal polyps. These methods allow identification of triggering allergen(s) and subsequent institution of appropriate therapy, i.e., allergen avoidance and/or desensitization treatments.

(Choice 2) is necessary only when nasal polyps have not regressed with pharmacologic therapies, namely topical application of steroids (beclomethasone) or a short course of oral prednisone. Histopathologic examination of removed nasal polyps usually confirms the allergic, inflammatory nature of the process by demonstrating chronic inflammatory infiltration and tissue eosinophilia.

(Choice 3) is a direct allergen challenge performed by inhalation of allergens through the nose. This test may allow identification of involved allergens in case of a positive reaction.

76. Question

1 points

Category: Pediatrics

A 3-year-old girl presents to the pediatrician's office. The mother states that the girl has been having big, bulky stools that float in the toilet. She also has intermittent diarrhea. On examination, her height is 88 cm (34.6 in, <5th percentile) and weight is 15.8 kg (34.8 lb, <5th percentile). In addition, she has an uncle who died of recurrent lower respiratory infections. Which of the following would be most effective for alleviating the gastrointestinal symptoms of this patient?

1. ☐ Avoidance of dairy products
2. ☐ Elimination of dietary fat
3. ☐ Ketogenic diet
4. ☐ Oral metronidazole
5. ☒ Pancreatic enzyme replacement ✓

INCORRECT ✗

The correct answer is 5.

This 3-year-old girl has steatorrhea, recurrent diarrhea, and failure to thrive. There is a family history of an uncle who died of recurrent lower respiratory infections. This patient most likely has cystic fibrosis (CF). The gastrointestinal system is significantly affected by CF. Gastrointestinal manifestations are caused by pancreatic exocrine insufficiency, which occurs in more than 85% of patients with CF. The mutation of the CF gene causes inspissation of pancreatic secretions and obstruction of the ductules. It causes the malabsorption of protein and fat, resulting in steatorrhea. Untreated pancreatic insufficiency results in malnutrition and failure to thrive. The treatment is to replace the missing pancreatic enzymes by oral supplementation of purified pancreatic enzymes (pancrelipase) with food. Several capsules are taken with each meal and snack. Even with pancreatic enzyme replacement, however, there might still be mild steatorrhea. Fat-soluble vitamins, such as A, D, E, and K, are also supplemented in these patients to avoid fat-soluble vitamin deficiency. Successful nutritional management is an important part of the overall management of CF, because good nutritional status improves the overall prognosis and decreases the frequency of pulmonary infections.

(Choice 1) is useful in diarrhea caused by lactose intolerance but not in CF.

(Choice 2) is inappropriate. In fact, the intake of fat should be encouraged because the fat content increases palatability and provides high calories to the patient's body. Dietary fat is also essential for the absorption of fat-soluble vitamins.

(Choice 3) has no role in the management of CF. It is sometimes used to control epilepsy.

(Choice 4) is effective against anaerobic bacterial infection. It can be useful in bacterial overgrowth in the intestine and in Clostridium difficile colitis, but it has no role in the treatment of pancreatic insufficiency.

77. Question

1 points

Category: Pediatrics

A 12-year-old boy with cystic fibrosis presents to the emergency department with a 3-day history of severe coughing, which is productive of yellow-greenish purulent sputum. He had fever and chills at home. He also complains of chest congestion and chest pain that is worse with coughing. On physical examination, his temperature is 39.6 °C (103.2 °F), blood pressure is 98/68 mm Hg, pulse is 102/min, and respirations are 24/min. He is noted to be lethargic. He has rales on the left lower lung field on auscultation, and chest radiography shows an infiltrate in the left lower lobe. Which of the following is the most appropriate initial antimicrobial therapy for this patient?

- ☐ Amoxicillin-clavulanate and gentamicin
- ☐ Azithromycin and ceftriaxone
- ☒ Ceftazidime and tobramycin ✓
- ☐ Levofloxacin and metronidazole
- ☐ Trimethoprim-sulfamethoxazole and vancomycin

INCORRECT ✗

The correct answer is 3.

Cystic fibrosis (CF) is one of the most common genetic diseases in the white population, with an incidence of about 1:3300. It is an autosomal-recessive disease caused by an genetic defect in the long arm of chromosome 7. CF affects mainly the pulmonary and the gastrointestinal systems. It causes recurrent, severe, lower respiratory tract infections leading to progressive pulmonary obstructive disease. Pneumonia is one of the major causes of morbidity and mortality in these patients. Pulmonary exacerbation of CF often manifests as fever, coughing productive of purulent sputum, retraction, tachypnea, shortness of breath, and chest congestion. The primary pathogen in children is *Pseudomonas aeruginosa*. Other pathogens include *Staphylococcus aureus*, *Haemophilus influenzae*, and gram-negative bacilli, such as *Escherichia coli*. The choice of initial antibiotic therapy is based on the above possible organisms. In addition, these patients have to be treated aggressively and promptly, because recurrent untreated episodes will result in bronchiectasis and significant decreases in pulmonary function. As a general rule, gram-negative organisms need to be covered with two antibiotics. In the case of *P. aeruginosa* infections, two different IV antibiotics that have antipseudomonal activity should be used. The most common choices are ceftazidime and tobramycin; an alternative combination is ticarcillin and tobramycin. Adjunctive therapies, such as chest physical therapy and bronchodilators, are also important.

(Choice 1) Amoxicillin-clavulanate is ineffective because resistance of *P. aeruginosa* is not secondary to the production of β -lactamase.

(Choice 2) Azithromycin is a macrolide that is not commonly used for *P. aeruginosa*, and ceftriaxone is not effective against *P. aeruginosa*.

(Choices 4 & 5) Levofloxacin, metronidazole, trimethoprim sulfamethoxazole, and vancomycin are not used to treat *P. aeruginosa*, but vancomycin can be useful if methicillin-resistant *S. aureus* is recovered from the sputum culture.

78. Question

1 points

Category: Pediatrics

A 2-weekpostmature neonate exhibits severe respiratory distress immediately after birth. Previously, green tinged meconium was noted in the amniotic fluid. Which of the following is the most appropriate next step in management?



1. ☐ Chest x-ray
2. ☒ Suctioning of the mouth and nasopharynx ✓
3. ☐ Oxygen supplementation by face mask
4. ☐ Intubation with mechanical ventilation
5. ☐ Emergency tracheostomy

INCORRECT ✗

The correct answer is 2.

Meconium is the fetal stool, which is mostly composed of desquamated cells from the gastrointestinal tract admixed with enough bile to give the soft stool a greenish color. A distressed fetus will pass meconium into the amniotic fluid and then may aspirate it. These infants often have placental insufficiency as a result of conditions such as maternal preclampsia, hypertension, or postmaturity. The aspirated meconium is very irritating to the lungs and causes a chemical pneumonitis. Postmature infants are particularly likely to have severe problems, because the meconium is diluted much less in their small amniotic fluid volume. The most important initial step in therapy is prompt suction of the nasopharynx and mouth to remove the meconium before more (or even any) is aspirated. This can be performed even before the infant is fully delivered, as soon as a head coated with meconium emerges from the birth canal.

(Choices 1 & 5) Neither chest x-ray nor emergency tracheostomy is warranted at this point.

(Choice 3) is deferred until after the nasopharynx and mouth are cleared of meconium.

(Choice 4) may become necessary if oxygen supplementation by face mask is unsuccessful.

79. Question

1 points

Category: Pediatrics

A 4-year-old girl is brought to the office due to seizures that occurred 2 hours ago. This is her first episode. Her parents are also concerned because she is not able to carry on activities that children of her age are expected to do. On examination, there is a red flat lesion covering the left eye area and adjacent facial skin, which does not blanch on pressure, and which the parents say has been present since birth. Other pertinent findings are hemianopia, hemiparesis, and hemisensory disturbances. An x-ray of the skull shows tramline intra-cranial calcifications. What is the most likely diagnosis?

1. ☒ Sturge-Weber syndrome ✓
2. ☐ Tuberous sclerosis
3. ☐ Capillary hemangioma
4. ☐ Epilepsy
5. ☐ Neurofibromatosis

INCORRECT ✗

The correct answer is 1.

The presentation of Sturge-Weber syndrome can include focal or generalized seizures, mental retardation, and a port wine stain or nevus flammeus along the territory of the trigeminal nerve, which represents a congenital unilateral cavernous hemangioma. Seizures are the usual neurological presentation and may start at any age. Other pertinent findings are hemianopia, hemiparesis, hemisensory disturbance, and ipsilateral glaucoma. Skull x-rays, taken after the age of 2 years, reveal gyriform intracranial calcifications that resemble a tramline.

Treatment is aimed at controlling the seizures and reducing intraocular pressure. Argon laser therapy is successful in removing the skin lesions.

(Choice 2) The initial presentation in tuberous sclerosis is also a seizure, but the cutaneous abnormality is called adenoma sebaceum, which appears between 5-10 years of age.

(Choice 3) Capillary hemangioma usually presents at birth or appears by 6 months of age. It consists of small capillaries and presents as an irregular, raised lesion that blanches on pressure. Approximately 75% regress by age 7.

(Choice 4) Epilepsy is difficult to diagnose with just a single episode of seizures.

(Choice 5) The cutaneous lesions of neurofibromatosis are axillary freckling and café-au-lait spots. The neurological presentation is dependent on the location and growth of the tumors.

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80. Question

1 points

Category: Pediatrics

A physician sees a 7-year-old girl who has been referred to him by the school nurse, who informs him that the child often has cessation of speech in mid-conversation, accompanied by a blank stare and flickering of the eyelids. These symptoms last approximately 10 seconds before she resumes regular activity. The physician suspects that the child has a seizure disorder and asks the child to hyperventilate. During hyperventilation, the aforementioned symptoms appear. Which of the following electroencephalogram patterns is most characteristic of this disorder?

1. ☐ Hypsarrhythmia
2. ☐ Interictal slow-spike waves
3. ☐ Centrotemporal spikes
4. ☒ Three-per-second spike-and-wave pattern ✓
5. ☐ Four to six-per-second spike-and-wave pattern

INCORRECT ✗

The correct answer is 4.

Absence, or petit mal, seizures cease as quickly as they commence. Motor activity stops abruptly, and patients develop a blank stare with fluttering of the eyelids. The seizures are brief (10–30 seconds), but may occur many times throughout the day. They end abruptly and have no postictal stage. Absence seizures can be triggered by hyperventilation. An electroencephalogram (EEG) shows a typical three-per-second spike-and-wave pattern.

(Choice 1) is seen on EEG in patients with infantile spasms.

(Choice 2) Interictal slow-spike waves on EEG are seen with Lennox-Gastaut syndrome.

(Choice 3) is the EEG finding diagnostic of benign partial epilepsy.

(Choice 5) A four to six-per-second irregular spike-and-wave pattern can be found on the EEG of a patient with juvenile myoclonic epilepsy (Janz syndrome).

81. Question

1 points

Category: Pediatrics

A one-year-old girl is brought to the emergency department (ED) by her mother due to a one-day history of fever and drowsiness. The child has been irritable since yesterday. On examination, she is hypothermic, lethargic and has nuchal rigidity. She flexes her hips when her neck is flexed. She appears septic, and large petechial and purpuric lesions are developing on her body. In the ED, she suddenly becomes hypotensive. Despite aggressive fluid and antibiotic resuscitation, the child dies. What will most likely be revealed as the cause of death during the autopsy of this child?



1. ☐ Myocarditis and heart failure
2. ☐ Acute respiratory distress syndrome
3. ☐ Bone marrow failure
4. ☒ Adrenal gland failure
5. ☐ Acute renal failure

INCORRECT ❌

The correct answer is 4.

In an infant with meningococemia, watch out for Waterhouse-Friderichsen syndrome, which is characterized by a sudden vasomotor collapse and skin rash (i.e., large purpuric lesions on the flanks) due to adrenal hemorrhage. Fulminant meningococemia can occur after a meningococcus infection, and approximately 10-20% of infants present with vasomotor collapse, large petechiae and purpuric lesions. The condition carries an almost 100% mortality.

(Choice 1) In meningococemia, myocarditis does not occur. It can affect the joints, skin and brain, but the heart is spared.

(Choice 2) Acute respiratory distress syndrome (ARDS) generally does not present in such a dramatic fashion. Rather, it presents with signs of hypoxia and infiltrates on chest x-rays. It is a gradual process where oxygenation becomes increasingly difficult. Most individuals require mechanical ventilation.

(Choice 3) The bone marrow is not affected by meningococcus. Individuals may show leukocytosis with a leftward shift.

(Choice 5) Meningococemia does not directly affect the kidneys. Renal failure may be secondary to sepsis and vasomotor failure, wherein severe vasomotor compromise results in decreased renal perfusion.

82. Question

1 points

Category: Pediatrics

A term, newborn male infant is being examined. He has a flat face, upward and slanted palpebral fissures, epicanthal folds, a simian crease, short and broad hands, high arched palate, and hypotonia. He was born to a 38-year-old Caucasian mother. What diagnosis is consistent with these findings?

1. ☒ Three copies of chromosome 21 ✓
2. ☐ Three copies of chromosome 18
3. ☐ Three copies of chromosome 13
4. ☐ Three copies of chromosome 8
5. ☐ Autosomal monosomy

INCORRECT ✗

The correct answer is 1.

The patient's manifestations (i.e., flat face, upward and slanted palpebral fissures, epicanthal folds, a simian crease, short and broad hands, high arched palate, and hypotonia) are characteristic for Down's syndrome (trisomy 21), which is the most common of all trisomies. This condition is usually caused by meiotic nondisjunction, and its likelihood increases with maternal age. Other manifestations include speckled irises (Brushfield spots), mental and growth retardation, pelvic dysplasia, hypoplasia of the middle phalanx of the 5th finger, intestinal atresia, high arched palate, and cardiac malformations. The diagnosis is confirmed by obtaining the patient's karyotype.

(Choice 2) Patients with trisomy 18 or Edward's syndrome typically present with low birth weight, closed fists (with the index finger overlapping the 3rd digit, and the 5th digit overlapping the 4th), narrow hips with limited abduction, short sternum, rocker-bottom feet, microcephaly, prominent occiput, micrognathia, cardiac and renal malformations, and mental retardation. It is much less common than Down's syndrome, and approximately 95% of patients die in their 1st year of life. This patient's manifestations are not characteristic of trisomy 18.

(Choice 3) Patients with trisomy 13 or Palau's syndrome typically present with cleft lip, flexed fingers with polydactyly, ocular hypotelorism, bulbous nose, low-set malformed ears, small abnormal skull, cerebral malformation, microphthalmia, cardiac malformations, scalp defects, hypoplastic or absent ribs, visceral and genital anomalies. None of these findings is present in the above case.

(Choice 4) Trisomy 8 is a very rare condition which presents with long face, high prominent forehead, wide upturned nose, thick everted lower lip, micro-retrognathia, low-set ears, high arched and sometimes cleft palate. Osteoarticular anomalies are common. You do not need to remember this one for the USMLE, as it has a very low chance of appearing (as the answer) in the exam.

(Choice 5) It is highly improbable (if not impossible) for this patient to have an autosomal monosomy because all complete autosomal monosomies are lethal early in development.

83. Question

1 points

Category: Pediatrics

A 7-month-old infant is brought to the emergency department. The mother states that the patient previously had been well but fell from a sofa onto the carpeted floor while taking a nap. The infant cried immediately according to the mother. However, on physical examination the child is noted to be lethargic. He is also noted to have retinal hemorrhages. Computed tomography of the head is performed and shows a subdural hematoma.

After initial stabilization of this patient, which of the following is the most appropriate next step in management?

1. ☐ Administer a pain reliever.
2. ☐ Notify the patient's primary care provider.
3. ☒ Contact the state agency that deals with child abuse. ✓
4. ☐ Obtain consent from the mother to admit the infant to the hospital.
5. ☐ Tell the parents that you suspect that the child has been abused.

INCORRECT ✗

The correct answer is 3.

Subdural hematoma and retinal hemorrhages are common signs of fatal child abuse. In this case, they are signs of shaken baby syndrome, in which a child is shaken to stop him or her from crying. Initially, the patient must be medically stabilized using the ABCs (airway, breathing, circulation) of resuscitation. The next step, if abuse is suspected, is to consult social services and the state agency that deals with this problem.

(Choices 1,2,4 & 5) Ongoing medical care and notification of the patient's primary care provider can be carried out after the state agency has been contacted. In cases of suspected child abuse, the physician neither has to inform the parents that child abuse is suspected nor obtain consent from the parent to admit the child to the hospital.

84. Question

1 points

Category: Pediatrics

A 6-year-old boy presents in clinic for a routine visit. Examination reveals coarse, dark pubic hair, an enlarged penis and testes, and acne of the face and upper back. His mother notes that he has a body odor similar to that of her teenage son after playing sports. The child is in the 99th percentile of height for his age group. Which of the following is the most likely diagnosis?

1. ☐ Congenital adrenal hyperplasia
2. ☒ Hypothalamic tumor ✓
3. ☐ Klinefelter syndrome
4. ☐ Male pseudohermaphroditism
5. ☐ XYY syndrome

INCORRECT ✗

The correct answer is 2.

The child is exhibiting isosexual precocious development. The enlarged gonads indicate that he has an increased exposure to gonadotropins, which may be the result of precocious release. Thus, increased testosterone is not the primary abnormality but is secondary to increased exposure to gonadotropins. In boys, a hypothalamic tumor can be found in an appreciable percentage of cases.

(Choice 1) An ectopic source of androgenic hormones, such as congenital adrenal hyperplasia, causes gonadotropins to be shut off and the testes to be small.

(Choice 3) is a common cause of primary hypogonadism.

(Choice 4) refers to infants 1 who are 46,XY males with testes but appear to have incomplete masculinization, including hypospadias. This can be the result of defects in testosterone synthesis, metabolism, or action at the cellular level.

(Choice 5) is associated with acne and tall stature but not with precocious puberty.

85. Question

Category: Pediatrics

1 points

A 29-year-old woman presents to the delivery room in labor at 35 weeks' gestation with a temperature of 40.0 C (104.0 F). She lives on a dairy farm and is in the habit of drinking unpasteurized milk from her cows before sending it to the dairy. For the past 3 days she has been unable to attend to her chores because of fever, headache, mild diarrhea, and a general feeling of illness. When her amniotic membranes rupture, the fluid is dark, cloudy, and brownish-green. At birth, the infant has no malformations or edema, but is in severe respiratory distress. Which of the following is the most likely diagnosis?

1. ☐ Congenital syphilis
2. ☐ Congenital toxoplasmosis
3. ☐ Fetal hydrops
4. ☐ Neonatal herpes
5. ☒ Neonatal listeriosis

INCORRECT ❌

The correct answer is 5.

Neonatal listeriosis presents soon after birth, usually within 5 days. The usual neonatal presentation is predominantly that of respiratory distress. Late-onset neonatal listeriosis is more likely to present with meningitis. Maternal infection is commonly acquired by contact with unpasteurized milk (mother works on a farm and drinks unprocessed milk), soft cheeses, deli meats such as hot dogs and pate, undercooked vegetables, and undercooked poultry. Typical symptoms in adults (exhibited by the mother) include fever, diarrhea, malaise, and a flu-like illness. Once maternal infection has been acquired, it can be transmitted to the fetus either vaginally or transplacentally. Amniotic fluid is often cloudy and discolored.

(Choice 1) Mothers with syphilis typically have a history of chancre or rash. Discolored amniotic fluid, acute fever and diarrheal illness are not typical. Severely affected infants may have pneumonia and hepatosplenomegaly.

(Choice 2) A symptomatic infant with congenital toxoplasmosis may display such signs as hydrocephalus, microcephaly, cataracts, hepatosplenomegaly, and a petechial and/or "blueberry muffin" rash. Respiratory distress is rare.

(Choice 3) is a nonspecific finding that can occur in a wide variety of conditions resulting in severe edema of the fetus. These include in utero heart failure, severe anemia, and intrauterine viral or spirochetal infection. In the present case, a typical maternal history compatible with listerial infection is obtained and no other blood or cardiac anomalies are present.

(Choice 4) can present with general collapse (associated with severe respiratory distress or liver failure), or as localized disease, including meningitis, ophthalmitis, or rash. The rash in infants is vesicular in its early stages, becoming crusted as the illness progresses. Many mothers do not have symptoms during their first infection; thus, a typical history of maternal genital herpes is not obtained in 70 to 90% of cases. However, the typical prodrome of listerial infection in the mother and lack of signs of neonatal herpes in either the mother or infant make this infection less likely.

86. Question

1 points

Category: Pediatrics

A 6-year-old girl is brought to the office due to significant steatorrhea and failure to thrive. She has a poor appetite and general malaise. Her past medical history is significant for prolonged neonatal jaundice and numerous respiratory tract infections. On examination, she looks a little underweight and has a runny nose. Auscultation reveals mild wheezing and diminished air entry in both lungs. Clubbing is visible on both hands. What is the best diagnostic test for this patient?

1. ☐ Blood cultures
2. ☐ Sputum cultures
3. ☐ CT abdomen
4. ☒ Sweat chloride test ✓
5. ☐ Liver function tests

INCORRECT ✗

The correct answer is 4.

In childhood, cystic fibrosis (CF) has a wide range of manifestations, such as anemia, heat intolerance, steatorrhea, wheezing, recurrent infections, hemoptysis, clubbing and rectal prolapse. In this child with failure to thrive, steatorrhea, and a history of neonatal jaundice, it is imperative to rule out CF. The steatorrhea of this child may be due to pancreatic insufficiency. A positive sweat chloride test is diagnostic, but it must be repeated twice to avoid any false positives or negatives. Genetic studies should be done when the sweat test is equivocal. Prenatal screening should be offered to high-risk families. The treatment is a high-calorie diet, pancreatic enzyme replacements and fat-soluble vitamins. The current median survival is 30 years. Complications of prolonged CF include gallstones, cirrhosis with portal hypertension and pancreatic fibrosis.

(Choice 1) Blood cultures cannot confirm the diagnosis of CF, but may be done as part of the work-up when sepsis is suspected.

(Choice 2) Sputum cultures are not diagnostic of CF, but allow for the identification of the organism responsible for the infections. The most common organisms are *S. aureus*, *Pseudomonas* and *H. influenza*. CF patients must be kept up to date with their immunizations.

(Choice 3) This patient's pancreatic insufficiency is most likely due to a lack of pancreatic enzymes. CT scan of the abdomen will generally not reveal any pathology.

(Choice 5) Liver enzymes are usually normal in CF patients. Furthermore, even if the enzymes are elevated, these are not diagnostic of CF.

87. Question

Category: Pediatrics

1 points

In the newborn nursery it is noted that a child has uneven gluteal folds. Physical examination of the child's hips reveals that one of them can be easily dislocated posteriorly with a jerk and a "click," and returned to normal position with a "snapping" sound. The family is concerned because a previous child had the same problem. Which of the following is the most likely diagnosis?

1. ☒ Developmental dysplasia of the hip ✓
2. ☐ Legg-Perthes disease (avascular necrosis of the femoral head)
3. ☐ Septic hip
4. ☐ Slipped capital femoral epiphysis
5. ☐ Traumatic delivery

INCORRECT ✗

The correct answer is 1.

What was formerly called congenital dislocation of the hip, and is now labeled "developmental dysplasia of the hip," runs in families and should be recognized at the time of birth by the physical examination maneuvers described in this vignette. In fact, a short-cut to pediatric hip pathology in the examination is to look at the age first. If the problem is noted on day one, this is the answer. Children are also prone to other hip problems, but the age of presentation is different for the alternative choices offered.

(Choice 2) Avascular necrosis is seen around 6 years of age, with pain and limping.

(Choice 3) follows febrile illnesses in toddlers who refuse to move the hip and do not let anyone try to manipulate it.

(Choice 4) affects chubby boys in their early teens that tend to limp and cannot rotate the hip internally.

(Choice 5) would not run in the family.

88. Question

1 points

Category: Pediatrics

A pediatrician performs a physical examination on a newborn he is asked to evaluate. The pertinent findings he notes include severe hypotonia, generalized weakness, and absent tendon stretch reflexes. The baby lies flaccid with little movement, but there appears to be preservation of the extraocular muscles. The patient is also observed to have fasciculations of the tongue, and the nurse informs the pediatrician that the infant has had trouble feeding. Which of the following is the most likely diagnosis?



1. ☐ Myotonic dystrophy
2. ☒ Werdnig-Hoffmann disease ✓
3. ☐ Infant botulism
4. ☐ Infantile myasthenia gravis
5. ☐ Duchenne's muscular dystrophy

INCORRECT ✖

The correct answer is 2.

Werdnig-Hoffmann disease is a spinal muscular atrophy caused by a pathologic continuation of a process of programmed cell death that normally occurs in embryonic life, but should cease at birth. It is inherited in an autosomal recessive fashion. Infants can be symptomatic at birth. Sparing of the extraocular muscles and sphincters is characteristic. Fasciculations are a sign of denervation of muscle and are best seen in the tongue. Infants assume a flaccid "frog-leg" posture. Most die by 2 years of age.

(Choice 1) A neonatal form of myotonic dystrophy appears in infants born to mothers with myotonic dystrophy. Clubfoot and contractures are common. Fasciculations are not seen.

(Choice 3) has a peak onset at 2–6 months of age. Sources for spores include honey, corn syrup, soil, and dust.

(Choice 4) can be transient as in those infants born of mothers with myasthenia gravis or, very rarely, a congenital manifestation of the disease itself. Fasciculations are not seen, and the extraocular muscles are not spared.

(Choice 5) is rarely symptomatic at birth.

89. Question

1 points

Category: Pediatrics

A 20-month-old male is brought to the emergency department for evaluation of rectal bleeding after his parents discovered a substantial amount of maroon colored stool when changing his diaper. He has no history of hematochezia. The patient has been otherwise healthy and has not appeared to be in any pain. He has been eating and drinking well. His temperature is 37 °C (98.6 °F), blood pressure is 85/50 mmHg, pulse is 130/min, and respiratory rate is 20/min. On examination, the abdomen is soft, nondistended, and nontender. On rectal examination, there are no fissures or masses present. A fecal occult blood test is positive. The remainder of the physical examination is normal. Which of the following is the best test to confirm the diagnosis?

1. ☐ Abdominal radiography
2. ☐ Superior mesenteric arteriography
3. ☐ Barium enema
4. ☒ Technetium-99m pertechnetate scan
5. ☐ Abdominal CT scan

INCORRECT ✖

The correct answer is 4.

Meckel's diverticulum is the most common congenital abnormality of the small intestine. It results from incomplete obliteration of the omphalomesenteric duct and usually contains ectopic pancreatic or gastric tissue. The most common age of presentation is 2 years old, and males are twice as likely to be affected as females. The majority of patients with Meckel's diverticulum are asymptomatic. In young children who are symptomatic, the most common presentation is painless rectal bleeding due to mucosal ulceration of the ectopic gastric tissue. The bleeding is often substantial, and children can present with severe anemia or hemorrhagic shock. Bleeding usually resolves spontaneously. The best diagnostic test is a technetium-99m pertechnetate scan (i.e. Meckel's scan), which detects the abnormal location of the gastric mucosa.

(Choice 1) Abdominal radiography is rarely helpful in diagnosing a Meckel's diverticulum because it cannot differentiate the diverticulum from loops of bowel.

(Choice 2) Superior mesenteric arteriography may be useful in detecting a Meckel's diverticulum, but it requires both the presence of a significant blood supply to the diverticulum and a steady flow of bleeding from the diverticulum. Since this test is more invasive and less reliable, a Meckel's scan would be a better first choice.

(Choice 3) A barium enema is diagnostic and often therapeutic in children with an intussusception, which is another cause of rectal bleeding in this age group. It is not useful in the diagnosis of a Meckel's diverticulum.

(Choice 5) An abdominal CT scan is not reliable because it often cannot differentiate a Meckel's diverticulum from loops of bowel.

90. Question

1 points

Category: Pediatrics

A concerned mother brings her 2-month-old daughter to the clinic because of constipation. The mother had appropriate prenatal care but decided to deliver her child at home with the help of a midwife. The child has not received any medical attention since birth. Examination reveals jaundice, an umbilical hernia, and poor muscle tone. Which of the following is the most appropriate diagnostic study?

1. ☐ Alpha-1-antitrypsin genotyping
2. ☐ Liver and spleen scan
3. ☒ Measurements of T₄ and TSH ✓
4. ☐ Barium swallow
5. ☐ RPR and FTA for syphilis

INCORRECT ✗

The correct answer is 3.

Measurements of T₄ and TSH levels are the most appropriate tests to confirm congenital hypothyroidism.

(Choice 1) is used to confirm alpha, antitrypsin deficiency.

(Choice 2) is useful to indicate absence of the biliary tree or blockage of the extrahepatic bile ducts and is therefore most useful to confirm biliary atresia.

(Choice 4) will indicate the presence of pyloric stenosis ("string" sign = narrowing of barium stream passing through the duodenum; "umbrella" sign = the hold-up of barium in the stomach).

(Choice 5) The RPR and FTA are serologic tests for syphilis.

91. Question

Category: Pediatrics

1 points

A 10-year-old girl is brought to the physician because of throat pain, anorexia, and fever for 2 days. Her temperature is 38.9 C (102 F). The patient's history is negative for allergic diseases. She has had two episodes of pharyngotonsillitis over the past several years. Examination reveals a purulent exudate in the posterior oropharynx and enlarged tonsils. There is bilateral tender enlargement of anterior cervical lymph nodes. Cardiac and chest auscultation is normal. A rapid strep test is positive. Which of the following is the most appropriate next step in management?

1. ☐ Confirmatory throat cultures before treatment
2. ☐ Symptomatic treatment with nonsteroidal anti-inflammatory drugs
3. ☒ Symptomatic treatment and oral penicillin V ✓
4. ☐ Symptomatic treatment and a broad-spectrum cephalosporin
5. ☐ Surgical referral for tonsillectomy

INCORRECT ✗

The correct answer is 3.

This child has an acute pharyngotonsillitis (sore throat). Most cases are caused by viruses, of which rhinovirus, coronavirus, and parainfluenza virus are the most frequent agents. Bacteria, and specifically group A β -hemolytic Streptococcus cause approximately 15 to 30% of cases. It is clinically important to determine whether pharyngitis is due to viruses or Streptococcus because of the different treatment involved. The classic symptomatology of "strep throat" is fever, anorexia, throat pain, and purulent exudate on the oropharyngeal mucosa.

Occasionally, there is an associated scarlatiniform rash. Clinical observation alone is not entirely reliable in differentiating between viral and streptococcal pharyngitis. Throat cultures or rapid strep test (the latter based on an enzyme immunoassay method that detects streptococcal antigens) help the clinician confirm or rule out streptococcal infection. Available rapid strep tests are highly sensitive (95%) but not very specific. Thus, a positive test confirms streptococcal infection and does not need confirmatory throat cultures before treatment

(Choice 1). If the rapid strep test is negative, on the other hand, cultures are necessary to rule out streptococcal infection. The treatment of choice is a full course of oral penicillin, combined with acetaminophen or anti-inflammatory drugs for symptomatic relief

(Choice 2) would not be adequate in this case because a rapid strep test has confirmed the streptococcal etiology. Antistreptococcal antibiotic therapy is mandatory to prevent complications (i.e., rheumatic fever).

(Choice 4) should be discouraged, since the indiscriminate use of broad-spectrum antibiotics facilitates the emergence of antibiotic resistance. Antistreptococcal treatment should be based on agents with narrow activity against group A Streptococcus (penicillin V or, in penicillin-allergic patients, erythromycin).

(Choice 5) is unnecessary in this case. Indications for tonsillectomy are rather limited, namely tonsillar enlargement causing persistent respiratory problems, swallowing difficulties, or obstructive sleep apnea.

92. Question

1 points

Category: Pediatrics

A 6-year-old girl is brought to the pediatrician because of severe dysuria. The history is unremarkable except for the patient having a sore throat. There is no history of hematuria, fever, use of bubble bath, pinworms, tight clothing, nylon underwear, or recent medication. Inspection of the perineum reveals an intense erysipelas-like erythema of the vulva, distal vagina, and perineal area. A serosanguineous vaginal discharge is present. Which of the following is the most likely cause?

1. ☐ Haemophilus influenzae
2. ☐ Neisseria gonorrhea
3. ☐ Chlamydia trachomatis
4. ☒ Group A β -hemolytic streptococcus
5. ☐ Streptococcus pneumoniae

INCORRECT ❌

The correct answer is 4.

Nonvenereal infectious vulvovaginitis is not uncommon in prepubertal patients. Poor perineal hygiene is the most common cause of nonspecific vulvovaginitis and accounts for 70% of all pediatric vulvovaginitis cases. Most of these cases are secondary to fecal contamination. Bacterial vulvovaginitis due to a primary respiratory or skin pathogen may also be found. Group A β -hemolytic streptococcus may be associated with streptococcal nasopharyngitis or scarlet fever, or may occur alone. Vulvovaginal symptoms of group A β -hemolytic streptococcal infection include severe perineal burning, dysuria, and erythema similar in appearance to erysipelas in the perineal area. Most patients will have a serosanguineous or grayish white vaginal discharge.

(Choices 1 & 5) Streptococcus pneumoniae and Haemophilus influenzae may cause a purulent vaginal discharge causing vulvovaginitis following, or in conjunction with, an upper respiratory infection.

(Choices 2 & 3) Sexual abuse should be suspected when Neisseria gonorrhea or Chlamydia trachomatis are found in a prepubertal child. In this case, there is no history of bubble bath, pinworms, tight clothing, nylon underwear, or recent medications, all of which have been implicated in vulvovaginitis. Likewise, although there is severe dysuria, there is no history of hematuria or fever, which makes a urinary tract infection less likely.

93. Question

1 points

Category: Pediatrics

A 7-year-old boy is brought to the clinic by his mother, who states that he has been complaining of abdominal pain for 2 to 3 days. He has been afebrile, with no vomiting or diarrhea. His mother states she brought him to the office today because she noticed a rash on his legs that is getting worse, and he is now complaining of knee pain. On examination, there are palpable purpuric lesions on both legs and buttocks. He has pain around his ankle and knee joints with minimal swelling, and no warmth or erythema. Which of the following is the most likely diagnosis?

1. ☐ Dermatomyositis
2. ☐ Gastroenteritis
3. ☒ Henoch-Schonlein purpura ✓
4. ☐ Juvenile rheumatoid arthritis
5. ☐ Kawasaki disease

INCORRECT ✗

The correct answer is 3.

Henoch-Schonlein purpura is a syndrome of small blood vessel vasculitis that presents as a triad of purpuric ("palpable purpura") rash, intermittent abdominal pain, and arthritis. The rash usually affects the lower extremities, buttocks, and lower back. The abdominal pain is crampy, and there are often guaiac positive stools. The arthritis can be transient, most commonly affecting the knees and ankles.

(Choice 1) is an inflammation of striated muscle and skin. Most patients have fatigue, weight loss, anorexia, and fever. There is progressive weakness of proximal muscles.

(Choice 2) would be characterized by vomiting and diarrhea, often accompanied by fever. Gastroenteritis would not be associated with a purpuric rash or arthritis.

(Choice 4) presents with polyarthritis. Fatigue, fever, weight loss, morning stiffness, and refusal to walk may be presenting complaints. It is unlikely to present with a purpuric rash and abdominal pain.

(Choice 5) is characterized by prolonged fever, bilateral nonpurulent conjunctivitis, cervical lymphadenopathy, fissuring of mucous membranes, desquamation of extremities, and truncal rash. None of these symptoms are present in this case.

94. Question

1 points

Category: Pediatrics

A young woman with phenylketonuria (PKU) who wants to become pregnant asks her physician if any special preparation is required before conception to ensure that her baby is healthy. Her physician tells her that women with PKU who are not on low phenylalanine diets have a higher risk of spontaneous abortion than the general population and often give birth to infants with mental retardation, microcephaly, and other congenital anomalies; high levels of phenylalanine seem to be the cause of these problems. Therefore, prospective mothers who have PKU should be placed on a low-phenylalanine diet before conception. Which of the following values represents the highest blood phenylalanine concentration that provides a high probability of avoiding untoward complications?

1. ☐ 1 mg/dL
2. ☒ 6 mg/dL ✓
3. ☐ 11 mg/dL
4. ☐ 16 mg/dL
5. ☐ 21 mg/dL

INCORRECT ✖

The correct answer is 2.

Phenylketonuria (PKU) is caused by a deficiency in phenylalanine hydroxylase, which has a prevalence of 1 case per 11,000 births. The normal pathway is disrupted, so phenylalanine and other metabolites accumulate and cause brain damage. Clinically, affected infants are blond, with fair skin and blue eyes. A mild eczematous rash disappears with age. Vomiting can be confused with pyloric stenosis. Mental retardation is usually severe. The infants have a musty, mousey odor of phenylacetic acid. Screening is recommended after 72 hours of age and after feeding of proteins. Early institution of a diet low in phenylalanine is essential in preventing brain damage. Women who have PKU and want to have children should go on a low-phenylalanine diet before conception and throughout pregnancy to reduce the risk of spontaneous abortion and birth defects. The blood phenylalanine levels should be kept below 6 mg/dL throughout pregnancy.

(Choice 1) Blood levels of 1 mg/dL are below the risk level but are not the highest safe level.

(Choices 3, 4 & 5) Pregnant women with PKU and phenylalanine levels above 6 mg/dL have a higher risk of spontaneous abortions. Infants born to such mothers may have mental retardation, microcephaly, and/or a congenital heart anomaly.

95. Question

1 points

Category: Pediatrics

A 2-year-old child attends day care at a local neighborhood nursery. The parents report that one of his favorite activities at day care is to play in the sandbox. The parents tell you that the child especially enjoys this activity when the owner's puppies are in the sandbox too. According to the parents, until recently, the patient has been in his usual state of good health; in addition, he has no significant past medical history and his immunizations are up to date. However, at this office visit, the patient has wheezing, hepatosplenomegaly, and prominent peripheral blood eosinophilia. Which of the following diagnoses is most likely in this child?

1. ☐ Enterobius vermicularis infestation
2. ☐ Eosinophilic lung disease
3. ☐ Ascariasis
4. ☒ Visceral larva migrans ✓
5. ☐ Strongyloidiasis

INCORRECT ✗

The correct answer is 4.

Visceral larva migrans is caused by infection with *Toxocara* larvae. It is most common in children 1–4 years of age, especially if they have pica or have a tendency to place their fingers in their mouth and have close contact with dogs or cats, because *Toxocara* are common parasites of both. Sandboxes are common areas for both pets and children. Symptoms include fever, hepatomegaly, wheezing, pulmonary disease, and eosinophilia.

(Choices 1 & 2) Eosinophilic lung disease, sometimes called Löffler syndrome, is a group of disorders; they are linked by the common findings of eosinophilia and pulmonary infiltrates. Anal pruritus and insomnia are the presenting complaints in *Enterobius vermicularis* infestation. Eosinophilia does not occur in most cases as tissue invasion is absent.

(Choice 3) may cause fever, urticaria, allergic symptoms, and granulomatous disease. Vomiting, abdominal distention, and abdominal pain may also be present.

(Choice 5) Pulmonary infections with *Strongyloides stercoralis* (strongyloidiasis) are usually mild and may pass unnoticed. However, in immunosuppressed and malnourished children, massive parasite invasion with *S. stercoralis* may cause generalized abdominal pain, fever, and shock due to gram-negative sepsis.

96. Question

1 points

Category: Pediatrics

A 1-month-old infant is seen in a well-baby clinic. The mother states that the baby is constipated and feeds poorly. On examination, he is jaundiced, exhibits poor muscle tone, and has a large posterior fontanelle and an umbilical hernia. He has gained only 300 g since discharge from the normal newborn nursery. Which of the following would be the most appropriate diagnostic study in this infant?

1. ☐ Alpha-1-antitrypsin deficiency
2. ☐ Barium swallow
3. ☐ Liver and spleen scan
4. ☒ Measurements of T4 and TSH levels
5. ☐ Muscle biopsy

INCORRECT ❌

The correct answer is 4.

Measurements of T4 and TSH levels are the most appropriate tests to confirm congenital hypothyroidism.

(Choice 1) Alpha-1-antitrypsin genotyping is used to confirm α -1-antitrypsin deficiency.

(Choice 2) will indicate the presence of pyloric stenosis ("string" sign = narrowing of barium stream passing through the duodenum; "umbrella" sign = the holdup of barium in the stomach).

(Choice 3) is useful to indicate the absence of the biliary tree or blockage of the extrahepatic bile ducts and is therefore most useful to confirm biliary atresia.

(Choice 5) is used to diagnose myopathic conditions, including Werdnig-Hoffmann disease, muscular dystrophy, and metabolic inclusion diseases.

97. Question

1 points

Category: Pediatrics

A 10-year-old girl is evaluated by a pediatrician. She is already 5'8", tall and is taller than other members of her family were at this age. Her arms are disproportionately long compared with her trunk, and her sternum is outwardly displaced. Her joints are hyperextensible, particularly at the knees. Ocular examination demonstrates dislocation of one lens. Based on the most likely diagnosis, which of the following conditions is the most appropriate screening method to prevent morbidity?

1. ☐ Bone density
2. ☐ CT scan of the pituitary stalk
3. ☐ CT scan of the pituitary stalk
4. ☒ Echocardiography ✓
5. ☐ Electrocardiogram

INCORRECT ✗

The correct answer is 4.

Marfan syndrome is a connective tissue disease with an autosomal-dominant inheritance. Cardinal features of the disorder include tall stature, ectopia lentis, mitral valve prolapse, aortic root dilatation, and aortic dissection. Because cardiovascular disease (aortic dilatation and dissection) is the major cause of morbidity and mortality in these patients, early intervention with β -adrenergic blocking agents and careful echocardiography screening may help slow aortic dilatation and allow prophylactic aortic root replacement before dissection occurs. The average lifespan of an affected individual is reduced by 50% but is improving because of better recognition and management of cardiovascular abnormalities.

(Choice 1) is performed in conditions predisposing to secondary osteoporosis, including Cushing syndrome, thyrotoxicosis, hyperparathyroidism, malabsorption syndrome, and long-term steroid use. However, osteoporosis is not associated with Marfan syndrome.

(Choice 2) is appropriate when evaluating for a pituitary tumor as the source of this child's tall stature. It may also help in identifying a cause of precocious puberty. Most commonly, these pituitary tumors are growth-hormone secreting tumors. However, this patient presents with the characteristic features of Marfan syndrome.

(Choice 3) is the screening and confirmatory test of Cushing syndrome. Patients with Marfan syndrome are not at risk for Cushing syndrome.

(Choice 5) can identify changes in rhythm and electrical activity, ischemia, and muscular hypertrophy. However, it is not a good method of evaluating the risk for aortic root dilatation and dissection in Marfan syndrome patients.

98. Question

1 points

Category: Pediatrics

A 16-year-old girl has had a fever, vomiting, and watery diarrhea for the past 24 hours. She also complains of intermittent abdominal pain and generalized myalgia. On examination, she is slightly lethargic. Her temperature is 39.4 C (103 F), blood pressure is 75/50 mm Hg, and pulse is 150/min. Her conjunctivae and pharynx are hyperemic. She has a generalized erythematous maculopapular rash that spares the wrists. Which of the following will be the most appropriate treatment?

1. ☐ Amantadine
2. ☐ Gentamicin
3. ☐ Ketoconazole
4. ☒ Nafcillin ✓
5. ☐ Prednisone

INCORRECT ✗

The correct answer is 4.

The clinical presentation is consistent with toxic shock syndrome, which is caused by *Staphylococcus aureus* and usually occurs in women using highly absorbent tampons. Toxic shock syndrome is caused by the release of a toxin from a *S. aureus* infection, and the condition is potentially fatal. The rash spares the wrists. Treatment includes controlling the symptoms of shock, removing any device from the vagina, and administering N antibiotics. A β -lactamase resistant anti staphylococcal antibiotic (e.g., nafcillin, oxacillin, and methicillin) should be used. Appropriate cultures should be obtained to verify the identity of the pathogen.

(Choice 1) would be used for a viral infection such as influenza.

(Choice 2) provides good coverage for gram-negative rods. However, this patient has infection by a gram-positive coccus.

(Choice 3) is an antifungal agent that would be used in the treatment of candidal infections.

(Choice 5) is a steroid and would be considered if the patient was going into anaphylaxis. In that case, epinephrine should be considered as well. If the patient is believed to be in shock from adrenal insufficiency, then stress dose steroids must be given.

99. Question

1 points

Category: Pediatrics

A 5-year-old girl is being evaluated for generalized swelling. Her blood pressure is 98/60 mm Hg. Her laboratory results show:

Creatinine: 0.7 mg/dL

Albumin: 1.6 g/dL

Cholesterol: 360 mg/dL

Triglycerides: 400 mg/dL

C3 complement: 120 mg/dL (normal, >80 mg/dL)

Antinuclear antibody: Negative

Urinalysis: 1 RBC/hpf protein 400 mg/dL

Which of the following is the most likely diagnosis?

1. ☐ Membranoproliferative glomerulonephritis
2. ☐ Membranous glomerulopathy
3. ☒ Minimal change disease ✓
4. ☐ Postinfectious acute glomerulonephritis
5. ☐ Systemic lupus erythematosus

INCORRECT ✗

The correct answer is 3.

Generalized swelling in a 5-year-old girl with proteinuria, hypoalbuminemia, and hyperlipidemia suggests nephrotic syndrome. In the absence of other pathology, minimal change disease is the most common etiology of primary nephrotic syndrome in childhood. It accounts for 80 to 85% of the cases of nephrotic syndrome. Minimal change disease has a peak occurrence between ages 2 and 8 years. In most of the cases, a viral illness precedes the proteinuria. The initial symptom is typically edema, which can happen around the eyes (periorbital edema) and in the feet (pretibial edema). Both blood pressure and renal function are normal in most patients. Hematuria is absent; complement C3 and C4 levels are normal. Immunosuppressive therapy with steroids is the mainstay of treatment. Minimal change disease is strikingly responsive to steroid therapy, with a 93% success rate with resolution of proteinuria. In cases that are resistant to steroids, cyclophosphamide may be used. In general, the prognosis for minimal change disease is excellent.

(Choice 1) is the most common cause of primary glomerulonephritis. It often presents in the 2nd decade of life with proteinuria, microscopic or gross hematuria, and hypertension. Renal function is usually normal. Hypocomplementemia is also commonly seen. Renal biopsy is diagnostic. The mainstay of treatment is alternate-day steroids.

(Choice 2) is very rare in the pediatric age group. Most cases are asymptomatic. It has been associated with various infections, the most frequent being hepatitis B infection. The diagnosis is confirmed by renal biopsy. The clinical course is very variable in children, but the overall prognosis is excellent, with a 50 to 60% spontaneous remission rate. Alternate-day steroid therapy may improve the prognosis.

(Choice 4) is a common cause of gross hematuria in children and usually develops 1-2 weeks after a streptococcal infection. Other organisms may also cause this disease. Clinical manifestations vary from asymptomatic microscopic hematuria to gross hematuria to acute renal failure. Tubular casts may be seen on microscopic examination of the urine specimen; C3 levels are decreased. The prognosis is generally good.

(Choice 5) is a systemic rheumatologic disease that usually presents with malar rash, photosensitivity, arthralgia, serositis, neurologic disorders, and immunologic abnormalities. It is associated with positive antinuclear antibodies and anti-double stranded DNA antibodies. Different forms of glomerulonephritis can occur in patients with this disease.

100. Question

1 points

Category: Pediatrics

A 38-year-old woman, gravida 1, para 0, delivers an appropriate-for-gestational-age infant by spontaneous vaginal delivery in the vertex position. The mother had excellent prenatal care and denies any history of tobacco, alcohol, or drug use. The infant weighs 8 lb 6 oz (3.8 kg). The Apgar score is 8 at 1 minute and 9 at 5 minutes. On physical examination, the infant is noted to have edematous swelling of the soft tissue of the scalp that crosses the midline. Which of the following is the most likely diagnosis?

1. ☐ Cephalohematoma
2. ☐ Subcutaneous fat necrosis
3. ☐ Fracture of the skull
4. ☒ Caput succedaneum 
5. ☐ Overriding of the parietal bones

INCORRECT

The correct answer is 4.

Caput succedaneum is a diffuse swelling of the soft tissues of the scalp. It extends across suture lines, and the edema starts to resolve over the first few days.

(Choices 1 & 3) Underlying skull fractures, not seen in caput succedaneum, are occasionally associated with cephalohematomas. The hemorrhages caused by cephalohematomas are subperiosteal; therefore, they are limited to one bone and do not cross suture lines. Cephalohematomas tend to increase in size over the first few days and resorb in approximately 3 months.

(Choice 2) Subcutaneous fat necrosis of facial or scalp tissue may be associated with a forceps delivery.

(Choice 5) Overriding of the parietal bones occurs when the bones of the scalp overlap at the suture line. The examiner would feel the overlap of bone, not a soft-tissue swelling.

101. Question

1 points

Category: Pediatrics

A 15-year-old Caucasian male is brought to the office by his mother for the evaluation of a six-month history of unstable gait and speech difficulty which are getting worse over time. His past medical history is insignificant. He is not taking any medications, and denies smoking or alcohol consumption. His blood pressure is 120/70 mmHg and pulse is 80/min. Musculoskeletal examination showed scoliosis and feet deformity with 'hammer toes.' The neurologic examination showed dysarthria, dysmetria, nystagmus, and absence of deep plantar reflexes on lower extremities. What is the most common cause of death in this patient population?

1. ☒ Cardiomyopathy ✓
2. ☐ Renal failure
3. ☐ Diabetes-related complications
4. ☐ Malignancy
5. ☐ Septic shock

INCORRECT ✗

The correct answer is 1.

This clinical scenario is typical for Friedreich ataxia, which is the most common type of spinocerebellar ataxias. It is an autosomal recessive disorder, and its symptoms usually begin before 22 years of age. Neurological manifestations (e.g., gait ataxia, frequent falling, and dysarthria) result from degeneration of the spinal tracts (spinocerebellar tracts, posterior columns, pyramidal tract). Non-neurological manifestations may also occur, and these include concentric hypertrophic cardiomyopathy, diabetes, and skeletal deformities (e.g., scoliosis and 'hammer toes'). The median survival after disease onset rarely exceeds 20 years. The most common causes of death are cardiomyopathy and respiratory complications. Cardiomyopathy develops in up to 90% of the patients.

(Choice 2) Renal failure is not characteristic of Friedreich ataxia.

(Choice 3) Diabetes develops in 10-20% of the patients with Friedreich ataxia, but it is a rare cause of death.

(Choice 4) Friedreich ataxia does not predispose to cancer.

(Choice 5) Septic shock may develop due to respiratory complications, but it is not as common as cardiac complications.

102. Question

1 points

Category: Pediatrics

A 15-month-old boy is brought into the clinic with a 1-day history of fever, decreased oral intake, and runny nose. His vital signs are within normal limits except for a temperature of 39.5 C (103.1 F). He is active and in no distress. His ears are clear bilaterally. There is a clear nasal discharge. Multiple small (1 to 2 mm) vesicular lesions are noted on the mucosa of the anterior tonsillar pillars and posterior palate. The rest of his examination is within normal limits. Which of the following is the most likely diagnosis?

1. ☐ Aphthous stomatitis
2. ☐ Hand-foot-and-mouth disease
3. ☒ Herpangina ✓
4. ☐ Kawasaki disease
5. ☐ Stevens-Johnson syndrome

INCORRECT ✗

The correct answer is 3.

Herpangina is a caused by coxsackievirus and usually manifests with dysphagia and vesicles on the anterior tonsils and palate. There is typically a history of fever and viral symptoms (runny nose, cough). Diagnosis is made by the presence of small vesicular lesions on the anterior tonsils and posterior palate.

(Choice 1) usually presents with ulcers on the buccal mucosa. There is no associated fever, and the patient does not usually complain of pain.

(Choice 2) is also caused by coxsackievirus and presents with small vesicles in the mouth, but there are also vesicles on the hands and feet. Commonly, a maculopapular rash is also present.

(Choice 4) is associated with a history of prolonged fever (more than 5 days), conjunctival erythema, cracked lips, maculopapular rash, cervical lymphadenopathy, and swollen hands.

(Choice 5) is a systemic disease that presents with ulcers over the entire mouth and lips. There usually is a confluent rash on the body as well. The child will appear very ill.

103. Question

1 points

Category: Pediatrics

A child is referred to a pediatric endocrinologist for further management of hypophosphatemia. On review of the family history, she notes that the patient's mother has fasting hypophosphatemia. Laboratory results show that the patient has a slightly reduced serum calcium level, a moderately reduced serum phosphate level, elevated alkaline phosphatase activity, and no evidence of secondary hyperparathyroidism. On physical examination, the child is noted to have smooth bowing of the lower extremities, coxa vera, genu varum, genu valgum, and short stature. Which of the following is another characteristic seen on physical examination in patients with familial hypophosphatemia?

1. ☐ Tetany
2. ☐ Rachitic rosary
3. ☐ Harrison's groove (pectus deformity)
4. ☐ Myopathy
5. ☒ Waddling gait ✓

INCORRECT ✗

The correct answer is 5.

Familial hypophosphatemia (also known as X-linked or as vitamin D resistant rickets) is the most common non-nutritional form of rickets. It is X-linked dominant, so some mothers as well as fathers may have clinical manifestations of the disease. Hypophosphatemia occurs because of defects in the proximal renal tubular reabsorption of phosphate caused by failure in the conversion of 25-(OH)-vitamin D to 1,25-(OH)₂-vitamin D. Bowing of the legs is a common presentation. In addition, these children may have a waddling gait and short stature. Calcium levels in the serum are normal, and thus there is no associated tetany, myopathy, or secondary hyperparathyroidism. Treatment consists of supplementation with phosphate and calcitriol. Two syndromes often confused with X-linked hypophosphatemia are vitamin D dependent rickets type I and receptor-deficient rickets (formally known as vitamin D deficient rickets type II). Neither consistently causes hypophosphatemia, and they are autosomal recessive conditions.

(Choices 1,2,3 & 4) Tetany, rachitic rosary, Harrison's groove (pectus deformity), and myopathy are present in calcium-deficient rickets.

Classic vitamin D deficiency rickets has become rare in the U.S. because of food supplementation and styles that bare much of the body to the sun.

104. Question

1 points

Category: Pediatrics

A 30-minute-old male newborn is noted to have central cyanosis. He was born to a 16-year-old white female at 28 weeks gestation. His respirations are rapid and shallow at 70/min. His other vital signs are stable. On examination, there is audible grunting, intercostal and subcostal retractions, nasal flaring, and duskiness. On auscultation, fine rales are heard over the lung bases. Nasal oxygenation does not improve his cyanosis. A chest roentgenogram shows fine reticular granularity, predominantly in the lower lobes. Arterial blood gas analysis shows evidence of hypoxemia and metabolic acidosis. What is the most likely diagnosis of this patient?

1. ☐ Transient tachypnea of the newborn
2. ☒ Hyaline membrane disease (HMO) ✓
3. ☐ Persistent pulmonary hypertension of the newborn (PPHN)
4. ☐ Meconium aspiration syndrome
5. ☐ Intracranial hemorrhage

INCORRECT ✗

The correct answer is 2.

Hyaline membrane disease (HMO) occurs primarily in premature infants. It occurs in 60-80% of infants who are less than 28 weeks gestational age. The decreased production and secretion of surfactant is the primary cause. In the absence of surfactant, atelectasis ensues and results in perfused but non-ventilated alveoli, and consequently hypoxia. HMO characteristically presents as tachypnea, prominent grunting, intercostal and subcostal retractions, nasal flaring, and duskiness within a few minutes after birth. Breath sounds may be normal or diminished with a harsh tubular quality, and on deep inspiration, fine rales may be heard, especially over the lung bases posteriorly. The characteristic chest-x ray finding demonstrates fine reticular granularity of the lung parenchyma. The clinical course, x-ray of the chest, blood gas and acid-base values help to establish the diagnosis. Treatment includes early mechanical ventilation and surfactant administration. In this neonate, the history of prematurity, the classic presentation, and the characteristic chest-x ray make HMO the most likely diagnosis.

(Choice 1) Transient tachypnea usually follows an uneventful normal term vaginal delivery or cesarean delivery. It is characterized by the early onset of tachypnea, sometimes with retractions or expiratory grunting, and typically, cyanosis that is relieved by minimal oxygen. The lungs are usually clear without rales or rhonchi, and the chest-x ray shows prominent pulmonary vascular markings, fluid lines in the fissures, over aeration, a flat diaphragm, and occasionally, pleural fluid. Hypoxemia, hypercapnia, and acidosis are uncommon. The distinguishing feature in this case is the x-ray finding.

(Choice 3) Persistent pulmonary hypertension of the newborn (PPHN) should be suspected in all term and post-term infants with cyanosis with or without fetal distress. Persistence of the fetal circulatory pattern of right-to-left shunting through the POA and foramen ovale after birth is due to very high pulmonary vascular resistance. The associated hypoxia is universal and unresponsive to 100% oxygen. The chest-x ray may be normal or may show parenchymal opacification in the chest, depending on the etiology. Although the clinical presentation of PPHN is indistinguishable from HMO, the x-ray findings of this case are very typical and therefore very suggestive of the latter.

(Choice 4) Meconium aspiration syndrome usually occurs in term or post-term infants. Either in utero or more often with the first breath, thick and particulate meconium is aspirated into the lungs, resulting in small airway obstruction and consequent respiratory distress that presents within the first hour of birth. Partial obstruction of some airways may lead to pneumothorax or pneumomediastinum. Patchy infiltrates, coarse streaking of both lung fields, increased anteroposterior diameter, and flattening of the diaphragm characterize the typical chest-x ray.

(Choice 5) Intracranial hemorrhage in neonates classically presents as periods of apnea, pallor or cyanosis, poor suckling, abnormal eye signs, high-pitched, shrill cry, muscular twitching, convulsions, decreased muscle tone or paralysis, metabolic acidosis, shock, and a decreased hematocrit. The fontanel may be tense and bulging. The condition is rarely present at birth and is diagnosed on the basis of the history, clinical manifestations, and trans-fontanel cranial ultrasonography or computed tomography (CT).

105. Question

1 points

Category: Pediatrics

A 2-week-old infant is noted to be jaundiced. The baby's stools are pale, and his urine is darkly colored. Physical examination demonstrates hepatomegaly. Serum studies show elevations of AST, ALT, conjugated bilirubin, and unconjugated bilirubin. By 2 months of age, the baby is notably irritated by pruritus, has retarded growth, and has visible dilated veins in the periumbilical area. Ultrasound fails to demonstrate a gallbladder. Which of the following is the most likely diagnosis?

1. ☐ Alpha-1-antitrypsin deficiency
2. ☒ Biliary atresia
3. ☐ Cystic fibrosis
4. ☐ Hepatitis B
5. ☐ Hepatitis C
6. ☐ Toxoplasmosis

INCORRECT ❌

The correct answer is 2.

The child has biliary atresia. This condition may either develop prenatally, in which case the gallbladder may be absent, or postnatally, probably after inflammation and scarring of the extrahepatic (and intrahepatic) biliary ducts. The underlying etiology is unclear. The history illustrated in the question stem is typical. In cases in which ultrasound is unrevealing, needle biopsy of the liver can yield tissue that usually excludes alternative diagnoses and can sometimes definitively demonstrate the atresia.

(Choices 1,3,4 & 5) Alpha-1-antitrypsin deficiency, cystic fibrosis, hepatitis B, and hepatitis C can all cause hepatitis in infancy but would not cause the gallbladder to be absent.

(Choice 6) Congenital toxoplasmosis results from a systemic infection with *Toxoplasma gondii*. When found in the newborn, congenital toxoplasmosis is always severe, and neurologic signs are always present. The classic triad is chorioretinitis, hydrocephalus, and intracranial calcifications. Other characteristic features include intrauterine growth retardation, jaundice, hepatomegaly, splenomegaly, lymphadenopathy, and rash.

106. Question

1 points

Category: Pediatrics

A premature infant born at 28 weeks' gestation develops respiratory distress shortly after birth. The infant is placed in an incubator with supplemental oxygen and is monitored in the neonatal intensive care unit. Given the severe prematurity of the infant, the physician gives strict instructions for the nursing staff regarding oxygen tension levels. Attempts at oxygen weaning are begun as soon as clinically feasible. Which of the following is characteristic of the eye damage that occurs following prolonged oxygen exposure in preterm neonates?

- ☒ Blood vessels in the vitreous ✓
- ☐ Cotton wool exudates in the retina
- ☐ Microaneurysm of the retinal arterioles
- ☐ Papilledema of the optic nerve head
- ☐ Ulcers on the cornea

INCORRECT ✗

The correct answer is 1.

The physician is trying to prevent retinopathy of prematurity, also known as retrolental fibroplasia. This is a cause of permanent blindness in premature neonates with respiratory distress syndrome who are treated with high oxygen levels. The pathophysiology of this condition is related to the fact that the premature infant's retinas are not yet fully vascularized, and the direction in which the vessels grow is partly in response to changes in oxygen tension. When the infant's eyes are not protected from increased environmental oxygen levels, the vitreous of the eye develops a higher oxygen tension than the retina, and the blood vessels grow abnormally into the vitreous (where they block the light) rather than staying on the retina. Shielding the neonate's eyes prevents this from happening and reduces the incidence of the complication of visual loss.

(Choice 2) indicate vascular leakage, which is not a feature of this condition.

(Choice 3) is relatively specific for diabetic retinopathy, which is seen in adults rather than in neonates.

(Choice 4) suggests increased intraorbital or intracranial pressure.

(Choice 5) is seen in gonorrhea ophthalmia and other serious conjunctival diseases.

107. Question

1 points

Category: Pediatrics

Approximately 19 days after having had a severe sore throat, a 10-year-old girl is taken to a pediatrician because she is complaining that her arms and legs hurt. The mother reports that before the extremity pain began, the child had a rash with irregular boundaries that lasted about a day. Physical examination demonstrates mild fever, as well as swelling and erythema around several large joints. Laboratory studies show an elevated erythrocyte sedimentation rate, and ECG demonstrates a prolonged PR interval. Which of the following is the most likely explanation for these findings?

1. ☒ Antigenic mimicry
2. ☐ Bacterial infection of valves
3. ☐ Parasitic infection of myocytes
4. ☐ Toxin production
5. ☐ Viral infection of myocytes

INCORRECT ✖

The correct answer is 1.

The child has acute rheumatic fever. Jones criteria for diagnosing rheumatic fever includes using both "major" criteria (carditis, polyarthritis, chorea, erythema marginatum, subcutaneous nodules) and "minor" criteria (arthralgia, fever, erythrocyte sedimentation rate elevation, C-reactive protein elevation, prolonged PR interval). Either two major criteria or one major plus two minor criteria can be used for diagnosis. Typically, by the time the acute rheumatic fever develops, the preceding streptococcal sore throat has healed, and the patient is no longer infected. The damage that is produced is immunologically mediated, based on antigenic mimicry of bacterial antigens for human ones. A number of specific targets have been proposed, including Lancefield group A antigen mimicking mitral valve glycoprotein, part of the M protein antigen mimicking helical proteins such as myosin, protoplast membrane mimicking myocardial sarcolemma or neuronal tissue, and bacterial hyaluronate capsule mimicking hyaluronate-containing articular tissues.

(Choice 2) Although streptococci, staphylococci, and other bacteria can infect cardiac valves, causing endocarditis, this would not be associated with arthritis and erythema marginatum.

(Choice 3) Parasitic + infection of myocytes can occur with either trichinosis or Chagas disease. Neither of these conditions would present with sore throat followed by arthritis.

(Choice 4) Toxic damage to the heart can occur with diphtheria but would accompany the sore throat, not be delayed by weeks.

(Choice 5) Viruses can cause myocarditis, but arthritis and erythema marginatum would not be present.

108. Question

1 points

Category: Pediatrics

A 2-year-old girl is brought to her pediatrician by her parents because of increasing lethargy and irritability. She has just started walking, and is teething and likes to chew on the woodwork around the windows. Physical examination reveals a tender abdomen. Laboratory studies indicate high iron and ferritin levels. The peripheral blood smear shows basophilic stippling. Which of the following is the most likely diagnosis?

1. ☐ Cobalamin deficiency
2. ☐ Diphyllobothriasis
3. ☐ Iron deficiency
4. ☒ Lead poisoning
5. ☐ Porphyria

INCORRECT ❌

The correct answer is 4.

Lead poisoning can occur if a child eats chips of old paint with lead in it. Lead inhibits ferrochelatase, an enzyme that catalyzes the insertion of iron into protoporphyrin, thus inhibiting the production of hemoglobin (the high iron levels reflect the inhibition of ferrochelatase). Basophilic stippling can be seen in thalassemia and in lead poisoning.

(Choice 1) can occur in pernicious anemia leading to megaloblastic anemia. Deficiency can result in neurologic symptoms of Paresthesia in the hands and feet, early loss of vibration and position sensation, and progressive weakness.

(Choice 2) is caused by *Diphyllobothrium latum*, a parasite that infects the jejunum and ileum, causing vitamin B₁₂ malabsorption and leading to megaloblastic anemia. Infection can cause abdominal discomfort and weakness. Praziquantel is highly effective in treating this infection.

(Choice 3) The desire to eat mud, paint, and ice is indicative of a condition called pica, which is suggestive of iron deficiency. Laboratory results would show elevated iron binding capacity and low ferritin level.

(Choice 5) In porphyria, heme biosynthesis is affected because of a defect in one of several enzymes involved in the construction of hemoglobin. The biochemical result of this condition is similar to that of lead toxicity, since lead affects ferrochelatase, an enzyme affected in some forms of porphyria. Children with porphyria can present with abdominal pain and sensitivity to sunlight.

109. Question

1 points

Category: Pediatrics

A mother brings her 6-year-old daughter for evaluations because she has never been able to toilet train her. The child states that she perceives the sensation of having to void, and empties her bladder normally at normal intervals, but is nonetheless wet with urine all the time. Which of the following is the most likely diagnosis?

1. ☒ Low implantation of one ureter ✓
2. ☐ Meatal stenosis
3. ☐ Ureteropelvic junction obstruction
4. ☐ Ureterovesical reflux
5. ☐ Urethral valves

INCORRECT ✗

The correct answer is 1.

One ureter is normally implanted into the bladder, and that one is responsible for the filling that leads to the normal voiding pattern. The other ureter has a low implantation, beyond the sphincter, into the vagina or the perineum. Having no sphincter, that ureter constantly leaks urine. Incidentally, this is a congenital anomaly that is symptomatic only in little girls. Boys have a longer sphincteric segment; even if a ureter has low implantation it does not leak.

(Choice 2) is a very common source of obstruction in little boys in the newborn period. It would not occur in little girls.

(Choice 3) is another common problem, but the clinical presentation is flank pain when urinary flow is much larger than usual. Typically this happens to adolescents when they drink a lot of beer for the first time in their lives.

(Choice 4) is also a common congenital anomaly, but the presentation is that of recurrent urinary tract infections.

(Choice 5) are seen in newborn boys who fail to void during the first day of life.

110. Question

1 points

Category: Pediatrics

An afebrile 12-week-old infant is brought to the pediatrician by his mother because of a persistent cough. Upon physical examination, the patient is found to have a temperature of 100°F (37.8°C) per rectum and is noted to have bilateral conjunctivitis. Other pertinent findings include tachypnea, inspiratory rales, and scattered expiratory wheezing. The rest of the physical examination is unremarkable. A complete blood count reveals eosinophilia. The chest x-ray film shows hyperinflation and patchy interstitial infiltrates bilaterally. Which of the following is the most likely diagnosis?

1. ☐ Respiratory syncytial viral pneumonia
2. ☐ Bronchiolitis
3. ☒ Chlamydial pneumonia ✓
4. ☐ Cystic fibrosis
5. ☐ The larval phase of ascariasis

INCORRECT ✗

The correct answer is 3.

Chlamydial pneumonia in infants characteristically presents at 3-16 weeks of age. Typically, the infants appear quite well, are afebrile, but have been ill with tachypnea and a repetitive staccato cough. Rales, and sometimes wheezing, can be heard. Conjunctivitis is present in approximately 50% of patients. Hyperinflation and patchy infiltrates are seen on chest x-ray films, and eosinophilia (>400 cells/mm³) is apparent upon a complete blood count (CBC).

(Choice 1) may present with temperature instability, respiratory distress, wheezing, apnea, clear nasal discharge, and poor feeding.

(Choice 2) causes inflammation of the bronchioles with narrowing of the bronchial diameter. It is a viral disease caused by RSV in 50% of cases, but also by Mycoplasma, parainfluenza viruses, adenoviruses, and other viruses. It manifests with low-grade fever, tachypnea, nasal flaring, rales, and expiratory wheezing. Chest radiography shows hyperinflation or atelectasis without infiltration.

(Choice 4) commonly presents as respiratory insufficiency with cough, dyspnea, bronchiectasis, and pulmonary fibrosis. The affected child also shows signs of malabsorption and failure to thrive.

(Choice 5) is asymptomatic. However, if a large parasite load penetrates the lungs, it may manifest with cough, hematemesis, eosinophilia, and pulmonary infiltrates.

111. Question

1 points

Category: Pediatrics

A one-month-old infant is brought to the physician for evaluation. His mother reports that for the past two weeks, he has been crying inconsolably for several hours every evening. His mother has tried multiple methods to calm the infant down, but nothing seems to work. The infant was born full term without complications. He takes two ounces of cow's milk-based formula every two hours and is growing well. His mother reports that he is happy and alert the rest of the day. The child's physical examination is unremarkable. Which of the following is a true statement about the child's condition?

- ☐ 1. The infant should be started on ranitidine for gastroesophageal reflux.
- ☐ 2. The infant should be changed to a lactose-free formula.
- ☐ 3. Bloody stools are often present.
- ☐ 4. The child will likely develop an anxiety disorder during childhood.
- ☒ 5. This condition usually resolves by four months of age. ✓

INCORRECT ✗

The correct answer is 5.

Infantile colic is defined as excessive crying in an otherwise healthy infant for more than three hours a day, more than three days a week, and more than three weeks in a month. Colic usually presents within the first three weeks of life and resolves spontaneously by four months of age. Most parents report that the crying occurs at the same time of day, typically in the early evening. The crying begins suddenly, and the child generally cannot be comforted. The cause of colic is not known. Some evidence suggests a digestive tract origin (e.g., trapped gas) leading to abdominal pain. Because the etiology of colic is not known, there is no universally accepted treatment. Proposed treatments include soothing measures, simethicone (which decreases the surface tension of gas bubbles), or probiotics.

(Choice 1) Gastroesophageal reflux usually presents with excessive spitting up and crying related to feedings. Medication is generally reserved for those infants not gaining weight appropriately.

(Choice 2) Changing to a lactose-free formula is not useful in infants with colic.

(Choice 3) Children with intussusception are usually 3-36 months in age. They have abdominal pain that occurs at different times of the day and bloody ("currant jelly") stools.

(Choice 4) There is no association between colic and anxiety disorders.

113. Question**1 points****Category: Pediatrics**

Five days ago, a 10-year-old child had an upper respiratory infection and was given symptomatic treatment and an antipyretic (i.e., aspirin for fever). She had seemingly recovered but now presents with fever, protracted vomiting, and lethargy. Physical examination reveals mild hepatomegaly. Serum analysis found total bilirubin, serum transaminases, and serum ammonia to be increased. Cerebrospinal fluid (CSF) was then obtained; it is normal, except for elevated pressure. Which of the following is the most likely diagnosis?

1. ☐ Hepatitis A virus (HAV)
2. ☐ Drug-induced hepatitis
3. ☒ Reye syndrome ✓
4. ☐ Infectious mononucleosis
5. ☐ Gilbert syndrome

INCORRECT ✗**The correct answer is 3.**

Reye syndrome is a secondary mitochondrial hepatopathy that, in genetically susceptible individuals, has a very high association with the ingestion of aspirin-containing medicines during influenza-like illnesses or varicella. The number of cases peaked in the 1970s, with a mortality rate of greater than 40%. Because the relationship between aspirin and Reye syndrome was discovered, aspirin is no longer recommended for treatment of children; consequently, the prevalence has since significantly decreased, but unless diagnosed and treated promptly the mortality rate has not changed. Reye syndrome has a biphasic course. It usually presents in a previously healthy child who had an upper respiratory infection or varicella. After the child seems to have recovered, the symptoms of vomiting, lethargy, and confusion appears and progress quickly. The patient may also be combative. Liver enzymes and the serum ammonia level are elevated. Treatment requires early recognition and control of increased intracranial pressure. Prognosis depends on duration of disordered cerebral function. Early diagnosis and treatment increases the likelihood for recovery to 90%.

(Choice 1) is an RNA-containing virus. It is a member of the Picornaviridae family and is spread predominantly by the fecal-oral route. The diagnosis of HAV infection should be considered when a history of jaundice exists in the patient's contacts, or if the patient or his or her family has traveled to an endemic area. The acute infection is diagnosed by the presence of immunoglobulin M (IgM) anti-HAV. Although elevation may be found in the bilirubin and serum transaminase levels, they do not help to differentiate the cause.

(Choice 2) is not the correct answer, because the patient in this scenario has a history of aspirin usage, and aspirin is excreted by the kidney.

(Choice 4) is a clinical syndrome caused by the Epstein-Barr virus (EBV). Clinical symptoms in children are often mild, but patients may have fatigue, fever, sore throat, and generalized lymphadenopathy. Splenomegaly may be prominent, causing left upper quadrant abdominal tenderness. Hepatomegaly may be present in a small number of patients. Symptomatic hepatitis or jaundice is uncommon. A mononuclear lymphocytosis with atypical-appearing lymphocytes is associated with infectious mononucleosis.

(Choice 5) is an inherited, benign, unconjugated hyperbilirubinemia that is transmitted as an autosomal dominant disease. The hyperbilirubinemia is mild, and the bilirubin level may fluctuate. Jaundice is intermittent and may be noted on routine physical examination. Results of liver biopsy and other liver function tests are normal.

114. Question

1 points

Category: Pediatrics

A neonate is markedly edematous and dies 1 hour after birth. A diagnosis of hydrops fetalis is made after the hematocrit on cord blood is demonstrated to be 5%. The erythrocytes in a smear from the cord blood are markedly hypochromatic. The mother is Rh positive and is known to have α -thalassemia trait. The thalassemia status of the father is unknown. Alpha-thalassemia is the suspected cause of the infant's hydrops. Which of the following hemoglobins would most likely be markedly elevated in this infant's blood if this diagnosis were correct?

1. ☒ Hb Barts ✓
2. ☐ Hbc
3. ☐ Hb Glower 2
4. ☐ HbH
5. ☐ HbS

INCORRECT ✗

The correct answer is 1.

The thalassemias are due to inadequate production of alpha or beta hemoglobin chains. The genes coding for the alpha chain have been duplicated, producing a total of four copies. If all four alpha genes are normal, or if three are normal and one is thalassemia, the person is clinically normal. If two are abnormal, the person has α -thalassemia trait with mild hypochromic anemia. If three are abnormal, the patient has HbH disease with hemolytic disease. If all four are abnormal, hydrops fetalis, clinically similar to that produced by Rh disease, is produced. Newborns with α -thalassemia trait, HbH disease, or hydrops fetalis produce Hb Barts, which corresponds to a tetramer of four gamma chains. Adults with HbH disease, and sometimes those with α -thalassemia trait, produce HbH, a tetramer of four beta chains.

(Choice 2) is an abnormal hemoglobin that, like HbS, can be associated with variable degrees of anemia.

(Choice 3) is a hemoglobin present in first trimester embryos.

(Choice 4) is seen in adults with α -thalassemia (three abnormal genes), but is made in infants.

(Choice 5) is associated with sickle cell anemia and trait.

115. Question

1 points

Category: Pediatrics

A 15-year-old boy is seen in the pediatrician's office for a health maintenance physical examination. The boy reports a heavy, dragging sensation in his left scrotum. The sensation is more pronounced after exercise. He denies any scrotal pain. He is not sexually active. Examination of his genitalia indicates Tanner stage 4. There is a palpable fullness over his left scrotum. Both testes are normal in size and smooth in contour. Which of the following is the most likely explanation of these findings?

1. ☐ Hydrocele
2. ☐ Inguinal hernia
3. ☐ Orchitis
4. ☐ Testicular tumor
5. ☒ Varicocele ✓

INCORRECT ✗

The correct answer is 5.

The findings in the history and physical examination are most consistent with the diagnosis of varicocele, which is an abnormally dilated pampiniform venous plexus. Varicocele is very common; it is found in 15% to 20% of male adolescents and is usually asymptomatic.

Patients with symptomatic varicocele usually describe a heavy, dragging sensation in the groin or the scrotum on the affected side.

Palpation along the spermatic cord often reveals the classic "bag of worms" mass above the testis. The size of the varicocele increases with Valsalva maneuver and decreases with lying down.

(Choice 1) is the presence of fluid within the tunica vaginalis. It may be noncommunicating, when the mass is confined to the scrotum, or communicating, when there is continuity from the tunica vaginalis to the peritoneum. Hydroceles are typically asymptomatic, although a large hydrocele may interfere with ambulation.

(Choice 2) usually presents with an increase in scrotal size. It occurs more commonly in males than in females. Surgical repair is indicated to prevent complications.

(Choice 3) is inflammation of the testis. It can be caused by sexually transmitted diseases, such as gonorrhea or chlamydia, or by viruses, such as Epstein-Barr virus, influenza, varicella, or coxsackie viruses. Orchitis usually presents with a painful and enlarged testicle.

(Choice 4) usually presents as a painless mass that is firm, hard, and inseparable from the testis.

116. Question

1 points

Category: Pediatrics

An infant is born with a craniofacial abnormality. There is a patch of very white skin on the midline forehead, above which is a white forelock of hair. The remainder of the hair is dark brown. The eyes are also abnormal, with color variations from brown to blue within the irises. The medial canthi are displaced laterally. The infant does not respond to sound. Which of the following is the most likely diagnosis?

1. ☐ Beckwith-Wiedemann syndrome
2. ☐ Fragile X syndrome
3. ☐ Goldenhar syndrome
4. ☐ Pierre Robin syndrome
5. ☐ Treacher Collins syndrome
6. ☒ Waardenburg syndrome ✓

INCORRECT ✗

The correct answer is 6.

The child most likely has Waardenburg syndrome, which is one of the more common craniofacial abnormalities. Defects of this type are related to maldevelopment of the first and second vertebral arches that form the face. The defects usually begin during the second month of gestation. The features illustrated in the question stem are typical. On a practical basis, you should remember that children with white forelocks (partial albinism) should be tested for hearing loss.

(Choice 1) is characterized by macroglossia, a large fontanelle, and a linear fissure in the external ear. Organomegaly of both the pancreas and kidney is common, and omphalocele may be seen at birth.

(Choice 2) Clinical features of fragile X syndrome include mental retardation, large testicles, a prominent jaw, large ears, and a large head. The syndrome is caused by a dominant X-linked trinucleotide repeat gene with only a 50% penetrance in females. Features are most prominent in affected males.

(Choice 3) produces an asymmetric face, malformed ears, small eyes, hearing loss, a large mouth, a small mandible, and vertebral anomalies.

(Choice 4) produces micrognathia, glossoptosis, and cleft palate.

(Choice 5) is characterized by malformation of the external ear, deafness, micrognathia, antimongoloid eye slanting, and coloboma of the lower eyelids.

117. Question

1 points

Category: Pediatrics

At a routine well-child visit, a mother reports that her infant is able to babble, coo, and form a social smile. When supine, the infant is able to follow an object 90 degrees from the midline through an arc of 180 degree. When prone, the infant is able to lift its head 45 degrees. The infant is also able to roll from the prone to the supine position. Which of the following is the most likely age of this infant?

- 1. ☐ Newborn
- 2. ☐ 1 month
- 3. ☐ 2 months
- 4. ☐ 3 months
- 5. ☒ 4 months

INCORRECT ❌

The correct answer is 5.

Rolling over from the prone to supine position usually does not occur until 4–6 months of age.

(Choices 1 & 2) From the age of newborn to 1 month, an infant may be able to regard a face, respond to a bell, and have equal movements.

(Choice 3) In addition, a 1-month-old may follow to midline. Babbling and cooing occur by 2 months of age. The social smile also is present by this time.

(Choice 4) Lifting the head and chest occurs by 3 months of age. A 2-month-old infant can use the eyes well enough to follow an object through 180 degrees.

Category: Pediatrics

A 3-year-old Caucasian boy of Scandinavian descent, who had been developing normally, contracted otitis media. Because he had not felt well, he missed two meals. Suddenly, he became lethargic, started vomiting, had a violent seizure, and became comatose. His parents called 911, and he was put on a saline IV drip and taken to the emergency department. There, his physician immediately ordered a complete blood count, a liver panel, a chemistry panel, and a urinalysis. A finger prick test showed that the child was hypoglycemic but not ketonic. In taking a history from the mother, the attending physician finds that an older sibling died mysteriously in her sleep as an infant. He immediately orders the saline IV drip to be changed to a 10% glucose solution, and shortly thereafter, the patient starts to recover. Several hours later, when the laboratory results become available, it is determined that the boy's serum glucose value had been 35 mg/dL and that he had slightly elevated serum blood urea nitrogen (BUN), ammonia, and uric acid levels: an anion gap metabolic acidosis; and his liver enzyme values were also elevated. However, both his serum and urine ketone levels were zero. The child most likely has which one of the following?

1. ☐ Glucose 6-phosphatase deficiency
2. ☒ Medium chain acyl-CoA deficiency
3. ☐ Carnitine acyltransferase deficiency
4. ☐ Primary hyperinsulinemia
5. ☐ Acute intermittent porphyria

INCORRECT ✖

The correct answer is 2.

This child almost certainly has medium chain acyl-CoA deficiency (MCAD). The first step in the β -oxidation of fatty acids is catalyzed by acyl-CoA dehydrogenases. There are at least four classes of acyl-CoA dehydrogenases: those that act on very-long-chain fatty acids (e.g., the 20-carbon arachidonic acid); those that act on long-chain fatty acids, C12 to C18; those that act on medium-chain fatty acids, C6 to C12; and, those acting on shorter-chain fatty acids, less than C6. Inborn errors inhibiting the activity of each of these dehydrogenases have been reported; these all are inherited as autosomal recessive traits, and all but medium chain acyl-CoA dehydrogenase (MCAD) deficiency are extremely rare. MCAD deficiency has a reported incidence of 1 per 6,500 births among North European Caucasians, and from 1 per 9,000 to 1 per 17,000 births in the general. Among certain ethnic groups, the presence of a mutant gene has been reported to be as high as 1 per 40 individuals. The prevalence of the disease is as at least as high as that of phenylketonuria, yet newborn screening is only required in a few states. The high prevalence rate has provoked a debate in many states concerning the need to require newborn screening. This has been inhibited by the unavailability of an inexpensive or readily available methodology: the classic method, tandem mass spectrophotometry requires a large investment in instrumentation, while molecular DNA probes are not yet available for such sizeable studies. Moreover, genetic analysis will only recognize the presence of the more prevalent K304E mutation. This MCAD gene is located on chromosome 1p31, and 26 different mutant forms have been characterized. However, studies conducted to date suggest the K304E MCAD mutation accounts for 90% of cases; 81% of these cases are homozygous for this gene, and 18% are compound heterozygotes. As in the case described, typically, the deficiency is characterized by sudden attacks after an infection and/or after fasting for 8–16 hours. The pathophysiology results from an inability to accomplish the first step in β -oxidation of mid-chain fatty acids, either those ingested (minor contribution) or longer-chain fatty acids that have been partially metabolized to 10–12 carbons and thus become the normal substrates for MCAD. Attacks are provoked by any situation that normally requires increased fatty acid oxidation, such as fasting or an illness. Since increased fat metabolism is not possible, continued existence in MCAD patients depends entirely upon glucose metabolism; consequently, hypoglycemia results. To make matters worse, gluconeogenesis is inhibited because acetyl-CoA cannot be formed but is required for the activity of pyruvate carboxylase to produce oxaloacetate from pyruvate; thus, amino acids cannot be used as a fuel. Furthermore, since acetyl-CoA cannot be formed, ketone bodies also cannot be synthesized and the brain is also deprived of this alternate fuel. The final insult is provided by the cell's attempt to metabolize the accumulated fatty acids by an endoplasmic reticular detoxifying P450 cytochrome mixed oxidase system that converts them into shorter-chain dicarboxylic acids by sequentially oxidizing and then removing the w-carbon by w-oxidation. This results in the accumulation of several unusual and abnormal mono and dicarboxylic acids and causes an anion-gap metabolic acidosis that will increase as more of these acids accumulate. Octanoic acid is one of these abnormal acids; it is a mitochondrial toxin suspected of inhibiting the urea cycle, causing an elevated blood urea nitrogen (BUN) level and hyperammonemia; the latter may induce an encephalopathy.

About 25% of babies succumb to respiratory or cardiac arrest during the first attack, which usually occurs between the ages of 6–24 months. Subsequent attacks are seldom fatal but are characterized by vomiting, seizures, lethargy, and sometimes coma and encephalopathy. Over the long term, the majority of individuals with identified symptoms have developmental and behavioral problems, often with chronic muscle weakness, attention deficit disorder, and/or cerebral palsy; however, others may seem normal. The penetrance is unknown, and it is believed that an unknown number of homozygous individuals never develop symptoms. Because infants who die during the first attack generate few physical clues relating to the cause of death, it was postulated that such cases are a common cause of the sudden infant death syndrome (SIDS). However, subsequent studies have shown that only 0.01% of SIDS cases are due to MCAD.

(Choice 1) Glucose 6-phosphatase deficiency, also known as type I glycogen storage disease or Von Gierke disease, also causes a severe fasting hypoglycemia and hepatomegaly, but the latter is due to accumulated glycogen, whereas in MCAT all carbohydrate stores are depleted. Carnitine acyltransferases I and II are required to shuttle fatty acids in and out of the mitochondria.

(Choice 3) As a consequence, carnitine acyltransferase deficiency results in an inability to use long-chain fatty acids as fuel, causing myoglobinemia and weakness following exercise.

(Choice 4) Although hypoglycemia occurs in hyperinsulinemia, a metabolic sequence similar to that described is not caused by fasting or illness.

(Choice 5) Acute intermittent porphyria is characterized by unexplained abdominal crises, acute peripheral or central nervous system dysfunctions, recurrent psychiatric illness, hyponatremia, and porphobilinogen in the urine during an attack.

119. Question

1 points

Category: Pediatrics

A 2-year-old boy presents to the emergency department with fever, irritability, and a skin rash 5 days after the onset of an upper respiratory infection. On examination, his temperature is 39.8 C (103.6 F), and his pulse is 94/min. There is an erythematous skin rash that involves his face, chest, back, and upper extremities. His skin is very tender to touch. Rubbing the skin causes separation of the epidermal layer. Which of the following is the most likely diagnosis?

1. ☐ Kawasaki disease
2. ☒ Staphylococcal scalded skin syndrome ✓
3. ☐ Streptococcal scarlet fever
4. ☐ Toxic epidermal necrolysis
5. ☐ Toxic shock syndrome

INCORRECT ✗

The correct answer is 2.

The boy in this clinical vignette has staphylococcal scalded skin syndrome (SSSS). It is characterized by fever, malaise, and a generalized fine erythematous rash. It is then followed by crusty, sometimes exudative, lesions around the mouth and eyes. It is usually preceded by an upper respiratory infection. The skin is tender to touch. With gentle rubbing, the epidermal layer can exfoliate, which is the characteristic Nikolsky sign. This syndrome is caused by an exfoliative toxin secreted by certain strains (SS and 71) of *Staphylococcus aureus*.

Sometimes, injection of the conjunctiva and fissuring of the lips can happen and might be confused with Kawasaki disease. The differential diagnosis includes toxic shock syndrome, scarlet fever, and toxic epidermal necrolysis. Nafcillin or oxacillin can be used to treat SSSS.

(Choice 1) involves vasculitis of epicardial coronary vessels, with possible development of aneurysm. The clinical syndrome presents with fever longer than 5 days, bilateral conjunctival injection, mucosal change in the oral pharynx, erythema of hands and feet with desquamation, and cervical lymphadenopathy. Transient arthritis may also occur.

(Choice 3) usually presents with an erythematous sandpaper-like rash with fever and "strawberry tongue." It is caused by erythrogenic exotoxins secreted by group A *Streptococcus*. It is usually associated with streptococcal pharyngitis.

(Choice 4) is a dermatologic emergency characterized by widespread epidermal necrosis and desquamation after formation of blisters and bullae. It is caused by a severe hypersensitivity reaction, usually to a drug. Major complications include sepsis, dehydration, and shock. Patients should be treated as if they have a generalized severe burn.

(Choice 5) is caused by an exotoxin (TSST-1) of *Staphylococcus aureus*, or less commonly, *Streptococcus pyogenes*. Patients present with high fever, myalgia, pharyngitis, abdominal pain, vomiting, diarrhea, headache, and a diffuse, sunburn-like rash. Severe cases are complicated by hypotension, shock, and renal failure.

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1 points

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120. Question

1 points

Category: Pediatrics

A 14-year-old black male comes to the office for the evaluation of pain in his right hip that started several weeks ago. The pain has gradually progressed, and now it limits his daily activities. He has sickle cell disease and was hospitalized three months ago due to a painful crisis that was successfully treated with hydration, oxygen, and analgesics. His temperature is 37.2 °C (99 °F), blood pressure is 100/70 mmHg, pulse is 80/min, and respirations are 16/min. Physical examination reveals no local tenderness, but there is restriction of abduction and internal rotation of the hip. What is the most likely diagnosis?

1. ☐ Osteomyelitis caused by Salmonella
2. ☐ Osteomyelitis caused by Staphylococcus
3. ☐ Joint effusion from septic arthritis
4. ☒ Avascular necrosis ✓
5. ☐ Femoral fracture

INCORRECT ✗

The correct answer is 4.

The most probable cause of this patient's complaints is aseptic necrosis of the femoral head. Pain in the hip that gradually progresses is a common clinical presentation. This disabling complication of sickle cell disease is relatively common, and has also been described with other hemoglobinopathies, such as SC hemoglobin disease. Other non-traumatic causes include chronic corticosteroid therapy, SLE, alcoholism, and Gaucher's disease.

The pathology involves occlusion of end arteries supplying the femoral head, bone necrosis, and eventual collapse of the periaricular bone and cartilage. The femoral head has two main sources of blood supply – ascending arteries and the foveal artery, which lie within the ligamentum teres. The foveal artery is patent early in life, but may become obliterated in older patients. This explains why aseptic necrosis of the femoral head is uncommon in children.

(Choices 1 & 2) Osteomyelitis is usually accompanied by high fever, and typically has an acute onset.

(Choice 3) A joint effusion may develop in patients with sickle cell disease, but the characteristic local warmth and tenderness are not present in this patient. Furthermore, the typical features of septic arthritis (i.e., fever, severe restriction of movement in all directions, inflammatory signs, and bacteremia) are also not present in this patient.

(Choice 5) A femoral fracture is unlikely without a history of trauma and with a gradually progressive course.

121. Question

1 points

Category: Pediatrics

A 1-year-old girl is brought to the emergency department by her mother because the child's "eyes and feet are dancing." On physical examination, the girl is well developed and in no acute distress. Her temperature 37.0 C (98.6 F), blood pressure is 100/55 mm Hg, pulse is 100/min, and respirations are 20/min. The patient has opsochlorus, myoclonus, and ptosis of the right eye. On history, the mother notes the child was born "looking like a blueberry muffin" and has had a persistent cough since the age of 2 months. Which of the following is the most likely diagnosis?

1. ☐ Astrocytoma
2. ☐ Glioblastoma multiforme
3. ☐ Hyperthyroidism
4. ☒ Neuroblastoma
5. ☐ Wilms tumor

INCORRECT ❌

The correct answer is 4.

Neuroblastoma arises from neural crest tissue and occurs in infancy. It is the most common solid tumor in children other than brain tumors. Eighty-five percent of neuroblastoma cases are diagnosed in children younger than 2 years. The clinical presentation varies with the tumor's location. Common signs and symptoms include asymptomatic abdominal mass, Horner syndrome, persistent cough, superior vena cava syndrome, bone pain, cord compression, subcutaneous nodules ("blueberry muffin" lesions), hypertension, opsochlorus ("dancing eyes"), and myoclonus ("dancing feet").

(Choices 1 & 2) Astrocytoma and glioblastoma multiforme are intracranial tumors and are therefore incorrect. The typical presentation of a brain tumor is early morning headache and vomiting, ataxia, cranial nerve palsies, nystagmus, and papilledema.

(Choice 3) does not cause opsochlorus or myoclonus, although it can cause tremor. Other typical features in children are irritability, excessive appetite, weight loss, increased perspiration, tachycardia, and goiter. Of note, hyperthyroidism occurs most frequently in children aged 12-14 years; only 2% of patients present before 10 years of age.

(Choice 5) occurs in the metanephric cells in the kidney. It is not associated with nervous system involvement.

122. Question

1 points

Category: Pediatrics

A 17-year-old boy was in his usual state of health until approximately 1 month ago when he developed a painful lump above his right knee. The patient says that he has fatigue and tells you that the leg pain awakens him from sleep. He also states that the leg pain is so bad that he has developed a limp that seems to be worsening. Physical examination reveals a hard, warm, tender mass. A radiograph of the right femur shows an "onion-skin" appearance at the location of the mass. Which of the following is the most likely diagnosis?

1. ☐ Osteomyelitis
2. ☒ Ewing's sarcoma
3. ☐ Osteosarcoma
4. ☐ Chondrosarcoma
5. ☐ Eosinophilic granuloma

INCORRECT ❌

The correct answer is 2.

Ewing's sarcoma is primarily a disease of adolescence but is far less common than osteosarcoma and is as likely to originate in almost any bone on the body. X-ray films are likely to show an "onion-peel," but this is not pathognomonic; the definitive diagnosis depends upon histologic examination, which will show small round cells. Primitive neuroectodermal tumors (PNETs) show a histologic pattern similar to that of Ewing's sarcoma, respond similarly to chemotherapy, and both are characterized by a 11,12 chromosomal translocation. In spite of resection, chemotherapy, and irradiation, Ewing's sarcoma still has a 50% mortality rate.

(Choice 1) is an infection of the bone, with the femur and tibia most commonly affected. Radiographs are normal early in the course of osteomyelitis.

(Choice 3) is the most common malignant bone tumor in children. The diagnosis of osteosarcoma may be suspected from good-quality radiographs. In advanced cases of osteosarcoma, possible findings include cortical destruction, sclerosis, a "sunburst" pattern of new periosteal bone formation, and calcified soft-tissue extensions.

(Choice 4) is rare in children. It occurs most commonly in the pelvis. Metastasis is usually by local extension.

(Choice 5) presents as a painless or mildly painful swelling in the skull, long bones, ribs, pelvis, or vertebra. Radiographs show a lytic lesion with well-defined borders.

123. Question

1 points

Category: Pediatrics

A 10-year-old boy is brought by his parents to the emergency department for the evaluation of headaches, fever, chills, and a rash over his neck, chest, and axillae. The rash appeared today, and for the past two days the child was complaining of a sore throat. He has no history of allergies, and his immunizations are up-to-date. His blood pressure is 112/70 mmHg, pulse is 108/min, respirations are 20/min, and temperature is 38.3 °C (101 °F). Examination reveals an erythematous rash with a sandpaper-like texture, and which blanches with pressure. There is submandibular tender lymphadenopathy, and the throat is erythematous with gray-white exudates. What is the most likely diagnosis?

1. ☐ Kawasaki disease
2. ☐ Staphylococcal scalded skin syndrome
3. ☒ Scarlet fever ✓
4. ☐ Herpangina
5. ☐ Mononucleosis

INCORRECT ✗

The correct answer is 3.

The patient most likely has scarlet fever. Scarlet fever is caused by strains of Group A streptococcus that produce erythrogenic exotoxins. It has the same mode of transmission and age of distribution as streptococcal pharyngitis. The illness may follow a streptococcal pharyngitis, wound infections, burns, or streptococcal skin infection. It begins acutely after an incubation period of 1 to 7 days. Initial symptoms include fever, chills, toxicity, abdominal pain, and pharyngitis. The rash initially appears on the neck, axillae, and groin within 12 to 48 hours, and subsequently generalizes within 24 hours. The rash characteristically has a punctate or finely papular texture which is sometimes readily palpable; hence, the "sandpaper-like" description. The pharynx is typically erythematous, swollen and possibly covered with gray-white exudates. The area around the mouth appears pale in comparison with the extremely red cheeks, giving the appearance of "circumoral pallor." Towards the end of the first week, desquamation begins in the face, progresses down the trunk, and finally extends to the hands and feet. The treatment is Penicillin V (drug of choice). Erythromycin, clindamycin, and first generation cephalosporins are good alternatives for patients who are allergic to penicillin.

(Choice 1) Kawasaki disease is a probable differential diagnosis, due to the patient's lymphadenopathy, changes in the buccal mucosa (pharyngitis and strawberry tongue), and rash; however, to make this diagnosis, at least one additional criterion must be present. That is, there should either be changes in the peripheral extremities or bilateral conjunctival injection. In a real clinical setting, it may be difficult to distinguish scarlet fever from Kawasaki disease, especially in children who are carriers of Group A Streptococcus; however, a rapid response to penicillin therapy generally confirms the diagnosis of scarlet fever.

(Choice 2) Staphylococcal scalded skin is caused by exfoliative strains of *S. aureus*, and is characterized by the development of superficial flaccid bullae followed by an extensive exfoliation of the skin. It is most common in infancy, and rarely occurs beyond five years of age.

(Choice 4) Herpangina is a throat infection caused by enteroviruses, especially Coxsackie A. It is characterized by a high fever and a severe sore throat that may result in a complete inability to swallow, sometimes necessitating IV hydration. Ulcerative lesions are found on the palate, tonsils, and pharynx. Sometimes, these lesions appear on the palms and soles, in which case the infection is called hand-foot-mouth disease.

(Choice 5) Mononucleosis can also present with exudative pharyngitis and lymphadenopathy; however, the characteristic rash of this patient makes scarlet fever the more likely diagnosis.

124. Question

1 points

Category: Pediatrics

A 3-year-old boy is brought to the office by his father for the evaluation of delayed milestones. According to the father, the boy is also very irritable, and he is worried because his nephew has been recently diagnosed with Down's syndrome. The mother has a serious drinking problem, and was recently arrested for "driving under the influence." On examination, the child's head circumference is 30cm (11 Bin), height is 75cm (29.5in), and weight is 12kg (26lb). He has a long philtrum with a thin upper lip border. What is the most likely explanation for the findings in this child?

1. ☐ Fragile X syndrome
2. ☐ Fetal hydantoin syndrome
3. ☐ Down's syndrome
4. ☐ Familial mental retardation
5. ☒ Fetal alcohol syndrome ✓

INCORRECT ✗

The correct answer is 5.

Fetal alcohol syndrome is characterized by irritability, mild to moderate mental retardation, hypoplastic maxilla, long philtrum, thin upper lip border and microcephaly.

(Choice 1) Fragile X syndrome is characterized by profound to normal IQ with learning disabilities, generalized language disability, hyperactivity, short attention span, autism, large head, prominent jaw, large low set ears and Macroorchidism.

(Choice 2) Fetal hydantoin syndrome is characterized by hypoplastic nails, cleft palate, and vitamin K deficiency, which may result in bleeding.

(Choice 3) Down's syndrome is characterized by mental retardation, microcephaly, flat occiput, Simian crease, endocardial cushion defects, prominent tongue, epicanthal folds with slanting eyes, and Brush field's spots on the periphery of the iris.

(Choice 4) The term 'familial mental retardation' doesn't exist.

125. Question

1 points

Category: Pediatrics

A 16-year-old girl comes to the physician's office because she has not begun menstruating yet. Both her mother and an older sister started menstruation at age 12. She takes no medication and denies strenuous exercise or excessive dieting. Her height is at the 50th percentile for age; her weight is at the 60th percentile. Both her breast and pubic hair development are at Tanner stage 4. Pelvic ultrasonography reveals a normal uterus and ovaries. Which of the following is the most likely diagnosis?

1. ☐ Imperforate hymen
2. ☒ Physiologic pubertal delay
3. ☐ Prolactinoma
4. ☐ Testicular feminization syndrome
5. ☐ Turner syndrome

INCORRECT ❌

The correct answer is 2.

Primary amenorrhea is defined as the absence of menses by the age of 16, or 4 years after thelarche (the onset of breast development). The prevalence of primary amenorrhea is 1 % to 2%. In this country, the median age of menarche is 12.5 years. Most girls have menarche about 2 years after thelarche.

Primary amenorrhea is different from secondary amenorrhea, which is defined as the absence of menses for three menstrual cycles or a maximum of 6 months in women who previously had normal menstruation. Primary amenorrhea has a vast differential diagnosis and is usually divided into three categories: 1) outflow tract anomalies (e.g., imperforate hymen, transverse vaginal septum, vaginal agenesis, testicular feminization); 2) end-organ disorders (e.g., ovarian failure, gonadal agenesis with 46,X,Y chromosomes); and 3) central disorders (e.g., hypothalamic disorders, pituitary disorders).

The age of the adolescent girl in this scenario is consistent with primary amenorrhea. Her sexual maturation is progressing appropriately. It is most likely that she has physiologic pubertal delay because the pelvic ultrasonography is normal. She probably has not completed her pubertal development.

(Choice 1) does not allow egress of menses. Patients usually present with pelvic pain from the accumulation of menses and dilatation of the uterus. Physical examination reveals a bulging membrane across the introitus and a midline pelvic mass.

(Choice 3) can cause headache, decreased growth rate, galactorrhea, delayed puberty, primary or secondary amenorrhea, gynecomastia, and visual field defects.

(Choice 4) is caused by either dysfunction or total absence of the testosterone receptor and results in a phenotypic female with 46,X,Y chromosomes. Patients with testicular feminization will have normal breast development but lack pubic hair. Pelvic ultrasound would reveal agenesis of the vagina and uterus.

(Choice 5) In Turner syndrome (45,XO), the ovaries undergo rapid atresia; by puberty, there are no primordial oocytes.

126. Question

1 points

Category: Pediatrics

A 12-year-old girl presents with a 2-month history of vaginal discharge. She describes it as clear and states that it stains her underwear. She says that she hates boys and that "no way" has she ever had sex or even kissed a boy. She reports having had developing breasts for 2 years and thinks that her growth spurt was about a year ago. Genital findings include a pubic hair stage of Tanner III with no evidence of redness or irritation of the vulvovaginal area. A slight amount of odorless, clear mucus is seen. Microscopic examination of the mucus reveals epithelial cells and a few bacteria, but no white cells. The pH is between 3.5 and 4. Which of the following is the most appropriate next step in management?

1. ☐ No treatment, but the girl should be reassessed in a few months ✓
2. ☐ Advise the girl to discontinue all bubble baths and wipe herself front to back after voiding
3. ☐ Pelvic examination to obtain cultures for gonorrhea and Chlamydia
4. ☐ Clotrimazole cream to be applied once a day for 10 days
5. ☐ Sitz baths one or two times a day and 1 % hydrocortisone cream applications once a day for a week

INCORRECT ✗

The correct answer is 1.

Physiologic leukorrhea is the most likely diagnosis. It frequently occurs a few months prior to menarche. Menarche may occur at Tanner stage III, usually about 1 year following the peak height velocity. The onset of menarche seems imminent in this girl. The pH of vaginal fluid falls below 4 with the onset of puberty. Certain vaginal infections alter the pH. For example, both trichomoniasis and bacterial vaginosis are characterized by a pH of greater than 5. There is no evidence of abnormal vaginal discharge, either by amount, color, or odor. No inflammation of the perineum is evident.

(Choice 2) Discussion of hygiene and possible irritants is particularly appropriate for prepubertal girls who have symptoms of dysuria or perineal irritation from bubble baths. This does not describe this patient.

(Choice 3) There are no indications at this time to perform a pelvic examination. The girl does not have the signs or symptoms of abnormal discharge or vulvovaginitis. A history of sexual contact was not obtained and seems unlikely in this case.

(Choice 4) Clotrimazole, miconazole, terconazole, and butoconazole are all commonly used to treat candidal infections. Candida or monilial vaginitis is characterized by clumpy white discharge, described as cottage cheese in appearance. Microscopically, pseudohyphae are seen. None of these are present in this patient.

(Choice 5) Sitz baths and a mild strength hydrocortisone cream may be helpful in soothing an irritated or mildly inflamed vulva. These are sometimes recommended as treatment for types of dermatitis, such as contact dermatitis, that involve the perineum. This is not necessary in this patient.

127. Question

1 points

Category: Pediatrics

A 14-year-old boy has pain in his left leg. An x-ray shows a tumor and a biopsy reveals histopathologic features characteristic of neural origin. Which of the following is the most likely diagnosis?

1. ☐ Chondroblastoma
2. ☒ Ewing sarcoma
3. ☐ Neuroblastoma
4. ☐ Osteosarcoma
5. ☐ Rhabdomyosarcoma

INCORRECT ✖**The correct answer is 2.**

Ewing sarcoma (ES) is the second most common bone cancer in childhood and adolescence. It has a peak age incidence during adolescent years. It rarely occurs in blacks and is more prevalent in boys than girls. ES may have histopathologic features characteristic of neural differentiation; ES is thought to be a tumor of neural origin, whereas most bone tumors are thought to be of mesodermal origin. The tumor category of ES has recently been expanded to include more neural tumors, such as peripheral primitive neuroectodermal tumor (PNET). Like other bone tumors, ES is associated with a consistent chromosomal translocation, $t(11;22)(q24;q12)$, which occurs in about 90 to 95% of cases.

(Choice 1) is a very rare bone tumor that most often occurs in the pelvis. Histopathology reveals cartilaginous tumor tissue with malignant spindle cells.

(Choice 3) is a common solid tumor of childhood that typically occurs during the first few years of life. Histologically it is a small round cell tumor with characteristic Homer-Wright rosettes. It displays neural differentiation and therefore might initially be confused with ES, but it is not a bone tumor. It usually occurs in the neck, thorax, abdomen, or flank.

(Choice 4) is the most common bone cancer of childhood and adolescence. Like ES, osteosarcoma may involve the bones of the leg, especially the femur, and frequently metastasizes to the lungs. Osteosarcoma is known to be associated with certain genetic conditions, such as osteogenic imperfecta, and there is an increased occurrence of osteosarcoma in children with retinoblastoma. It occurs in all races. Pathologically, neural differentiation is not observed.

(Choice 5) is the most common soft tissue sarcoma of childhood. It does occur more frequently among males and whites, but it is usually found in the head and neck areas and the genitourinary tract. Like neuroblastoma and ES it is a small round cell tumor. It does not display neural differentiation. It may first be noticed after trauma as in this case, but there is no bony involvement.

128. Question**1 points****Category: Pediatrics**

A 12-year-old, previously healthy girl presents to her physician with a chief complaint of early morning headaches. She states that these headaches wake her up from sleep 2 to 3 days a week. She also complains of some vomiting associated with the headaches. The headaches have been getting progressively worse for the past 2 months. She denies any photophobia, dizziness, or blurred vision. There is no history of a recent respiratory infection, runny nose, or cough. There is no history of recent trauma. In the office, her vital signs are within normal limits. Her examination shows pupils that are equal, round, and reactive, with no maxillary or frontal sinus tenderness. Her tympanic membranes are clear and intact. Her neck is supple with full range of motion. Neurologic examination shows a positive Romberg sign. Which of the following tests would most likely confirm the diagnosis?

1. ☐ CT of the brain
2. ☒ MRI of the brain
3. ☐ Plain film of the skull
4. ☐ Sinus x-ray film
5. ☐ Spinal tap

INCORRECT**The correct answer is 2.**

The early morning headaches associated with vomiting and getting progressively worse, along with the presence of a positive Romberg sign, suggest the presence of a space-occupying lesion (a brain tumor). An MRI of the brain would be the best diagnostic test to rule out brain tumors, posterior fossa tumors, which are of clinical concern in children.

(Choice 1) would be a good first test in most cases, except when you are looking for a brain tumor in children because it may not show a posterior fossa tumor. For investigation of an intracranial bleed, a CT would be an appropriate test, although there is no recent history of trauma to suggest this diagnosis.

(Choice 3) A plain skull x-ray film would provide information regarding any fracture or lytic lesions, both unlikely in this example.

(Choice 4) Sinus films would be helpful in the diagnosis of sinusitis, which could relate to chronic headaches. However, this patient had vomiting with headaches, no sinus tenderness, and no history of chronic rhinitis, so sinusitis would be an unlikely diagnosis.

(Choice 5) A spinal tap would reveal increased intracranial pressure or an infection of the cerebrospinal fluid. However, it would not be the first test to do before ascertaining whether there was a space-occupying lesion in the brain.

129. Question

1 points

Category: Pediatrics

A previously well 3-year-old child presents to the clinic with marked erythema of the cheeks that appeared suddenly overnight. The rash has spread to involve the proximal arms, and it has a reticular erythematous maculopapular appearance. There are no known allergies, and the child has no prior illnesses and has not been exposed to any individuals who are ill. Vital signs are within normal limits. Cervical lymphadenopathy is noted on physical examination. Which of the following is the most likely cause of this child's rash?

1. ☐ Contact dermatitis
2. ☒ Fifth disease ✓
3. ☐ Measles
4. ☐ Roseola
5. ☐ Rubella

INCORRECT ✖

The correct answer is 2.

Fifth disease, or erythema infectiosum, is caused by infection with human parvovirus B 19. The rash of erythema infectiosum starts on the cheeks, giving a "slapped cheek" appearance. The rash may disappear and recur.

(Choice 1) presents with a history of contact with an allergen. The skin is inflamed and itchy, with weeping and vesiculations. The site of involvement is often suggestive, e.g., the ear lobes and nape of neck (from nickel in jewelry), wrist (watch straps, bracelets), and feet (dyes in socks, tanning agents, glues in shoes). Typically, the dermatitis does not spread because the reaction is limited to the areas of contact.

(Choice 3) The rash of measles begins on the head and spreads down the body. It is erythematous and maculopapular, becoming confluent with progression.

(Choice 4) is caused by human herpesvirus 6 and has an incubation period of 9 days. The rash is maculopapular and develops 3 to 4 days after fever.

(Choice 5) produces a maculopapular rash that has no prodrome in children. Lymphadenopathy develops. The rash starts on the face and then becomes generalized.

130. Question

1 points

Category: Pediatrics

A 15-year-old girl has a round, 1-cm cystic mass in the midline of her neck, at the level of the hyoid bone. The mass is deep to the skin and moves slightly when the patient swallows. When the mass is palpated at the same time that the tongue is pulled, there seems to be a connection between the two. The mass has been present for at least 10 years, but only recently bothered the patient because it became infected. Which of the following is the most likely diagnosis?

1. ☐ Branchial cleft cyst
2. ☐ Cystic hygroma
3. ☐ Epidermal inclusion cyst
4. ☐ Metastatic thyroid cancer
5. ☒ Thyroglossal duct cyst

INCORRECT ❌

The correct answer is 5.

The age, description, location, and "connection with the tongue" are classic for a thyroglossal duct cyst. The thyroid gland initially develops in close proximity to the tongue, and then descends into the neck via the thyroglossal duct, which is normally obliterated but occasionally persists as thyroglossal duct cysts or nodules.

(Choice 1) Branchial cleft cysts are seen in the lateral aspect of the neck, along the anterior edge of the sternocleidomastoid muscle, anywhere from in front of the tragus to near the base of the neck.

(Choice 2) is seen at the base of the neck, as a soft, fluid-filled mass that goes deep into the mediastinum.

(Choice 3) Epidermal inclusion cysts are attached to the skin not connected to the tongue. If this cyst had been present for 10 years, it would have caused problems long ago. Furthermore, its location at the midline and level of the hyoid would have been an incredible coincidence.

(Choice 4) Thyroid cancer, if primary, would have been lower in the neck, where the thyroid is located. If it were metastatic, it would have been in the lateral side of the neck at the jugular nodes. Also, although thyroid cancers are notorious for slow growth, a cancer would be larger than 1 cm in size after 10 years.

131. Question

1 points

Category: Pediatrics

A 2-year-old child has had red, weeping, crusted lesions of the face, scalp, diaper area, and extremities since about age 2 months, with multiple periods of exacerbation and improvement. Attempts to remove potentially irritating substances have not modified the course of the rashes. The child is noted to be constantly scratching and rubbing involved areas. There is a strong family history of hay fever and asthma. Which of the following is the most likely diagnosis?

1. ☒ Atopic dermatitis ✓
2. ☐ Cellulitis
3. ☐ Contact dermatitis
4. ☐ Lichen simplex chronicus
5. ☐ Seborrheic dermatitis

INCORRECT ✗

The correct answer is 1.

This is atopic dermatitis. The presentation illustrated is typical for young children. Secondary bacterial infections and regional lymphadenitis may complicate the clinical picture. The dermatitis often improves by age 3 or 4 years, but periodic exacerbations may continue to occur into adulthood. Older children and adults tend to have more localized lesions, typically with erythema and lichenification. Common sites of involvement in older children and adults include the antecubital and popliteal fossas, eyelids, neck, and wrists. Once the diagnosis is made, the physician must remember not to make the mistake of automatically attributing all subsequent skin lesions the children develop to the atopic dermatitis, since they remain vulnerable to all other dermatologic diseases as well.

(Choice 2) is a streptococcal, or less commonly staphylococcal, infection causing acute inflammation of subcutaneous tissues associated with local erythema, tenderness, frequently lymphangitis, and regional lymphadenopathy. The skin is usually warm, edematous, and erythematous, and may exhibit a "peau d' orange" (orange peel) appearance.

(Choice 3) can resemble atopic dermatitis, but the long history illustrated in the question stem would not be typical of this condition.

(Choice 4) is characterized by itchy, dry skin that may progress to well-demarcated, hyperpigmented, lichenified plaques of oval, irregular, or angular shape.

(Choice 5) is an inflammatory scaling disease, usually of the scalp and face that may initially resemble atopic dermatitis in infants. Unlike atopic dermatitis, however, it tends to remain confined to the head and scalp. (The diaper area may be initially involved, but this usually clears with time.)

132. Question**1 points****Category: Pediatrics**

A 7-year-old boy is brought to the physician because of recurrent headaches. The child feels nauseated before and during each attack, and derives some relief from lying down in a dark room. Noises, bright light, and fatigue seem to trigger the episodes. The child frequently complains of headaches at school, and his mother has been occasionally compelled to take him home. The mother is worried about the possibility of a serious illness. She reports that the child's father has similar headaches. The child's growth is normal, and a neurologic examination fails to reveal any abnormality. Which of the following is the most likely diagnosis?

1. ☐ Brain tumor
2. ☐ Cluster headache
3. ☐ Conduct disorder
4. ☒ Migraine
5. ☐ Tension headache

INCORRECT ❌

The correct answer is 4.

Migraine may affect children as well as adults. It is the most common cause of headaches in children. Migraine in children manifests with symptomatology similar to that observed in adult patients. Certain triggers are easily identified, such as loud noises, bright lights, stress, fatigue, caffeine-containing drinks, and some foods. Parent education on what is known about migraine etiology is essential in management. Treatment consists of the measures that the parents or patients themselves have found to provide relief, plus acetaminophen or a nonsteroidal antiinflammatory drug (NSAID) if such measures are insufficient. Ergotamine should be reserved for the most severe cases, and sumatriptan is not yet approved for use in children.

(Choice 1) is very unlikely in the presence of classic symptoms of migraine, as in this case. The manifestations of a brain tumor in children are usually accompanied by evidence of increased intracranial pressure.

(Choice 2) is extremely rare in childhood. It manifests as attacks of intense pain occurring at night and is often precipitated by alcohol consumption. Each attack lasts up to 2 hours and recurs daily for a period of up to 8 weeks. These periods are then followed by a headache-free interval that usually lasts 1 year.

(Choice 3) is a childhood behavioral disorder that is similar to antisocial behavior disorder in adults. The child displays persistent patterns of aggression to animals or people, destruction of property, stealing, and opposition to parental guidance.

(Choice 5) is the second most important cause of headache in children and is more common in adolescents. This form of headache manifests as a viselike tightness around the head and in the neck muscles. Tension headache is frequently a response to stress or fatigue.

133. Question

1 points

Category: Pediatrics

A previously healthy 4-year-old child is brought to the physician for a school physical for kindergarten. His mother has brought in his immunization record and reports that received all his immunizations prior to age 2. He has received only a yearly influenza vaccine since age 2. His physical examination is unremarkable. Which of the following immunizations should he be given during this visit?



1. ☐ Haemophilus influenza type B #4
2. ☐ Pneumococcal #3
3. ☒ Inactivated polio #4 ✓
4. ☐ Measles-mumps-rubella #1
5. ☐ Hepatitis B #3

INCORRECT ✗

The correct answer is 3.

By the time a child starts school, he should have received 3 doses of the hepatitis B vaccine, 3 doses of the rotavirus vaccine, 5 doses of the Diphtheria-Tetanus-acellular Pertussis (DTaP) vaccine, 4 doses of the Haemophilus influenza type b (Hib) vaccine, 4 doses of the pneumococcal conjugate vaccine (PCV), 4 doses of the inactivated polio vaccine (IPV), 2 doses of the measles-mumps-rubella (MMR) vaccine, 2 doses of the varicella vaccine, and 2 doses of the hepatitis A vaccine. At the 4-5 year old visit, children should receive their DTaP #5, IPV #4, MMR #2, and Varicella #2. Children should receive 3 doses of IPV prior to 18 months and then a fourth booster dose at 4-5 years.

(Choice 1) The Haemophilus influenza type B series consists of 4 vaccines and should be completed by 15 months of age.

(Choice 2) Healthy children should receive 4 doses of the pneumococcal conjugate vaccine prior to age 15 months.

(Choice 4) Measles-mumps-rubella and the Varicella series consists of 2 doses each, the first is given at 12-15 months and the second is given at 4-5 years.

(Choice 5) The hepatitis B vaccine series consists of 3 doses and should be completed prior to 18 months in children who are on schedule.

134. Question

1 points

Category: Pediatrics

A 3-month-old, previously well male infant presents to the emergency department in January with a 2-day history of clear rhinorrhea, low-grade fever, and poor appetite, but no cough. On physical examination, there are mild subcostal retractions, coarse breath sounds heard throughout the lung fields, and scattered expiratory wheezes. The child receives an intravenous fluid bolus in the emergency department and is admitted for observation. Which of the following is the most severe, life-threatening complication of this child's illness?

1. ☒ Apnea ✓
2. ☐ Congestive heart failure
3. ☐ Dehydration
4. ☐ Hypoxemia
5. ☐ Wheezing

INCORRECT ✗

The correct answer is 1.

Apnea can be a life-threatening complication of RSV infection in infants younger than 1 year. For this reason, infants with respiratory distress and either laboratory confirmation or a clinical picture suggestive of RSV infection are often admitted to the hospital for supportive care and monitoring.

(Choice 2) is not associated with RSV infection in a child with normal cardiac function.

(Choice 3) Infants are obligate nose-breathers and may have difficulty feeding because of nasal congestion. In the face of respiratory distress, infants have increased insensible fluid losses and are at risk for dehydration. Dehydration, however, is not the most severe risk for this child.

(Choice 4) Hypoxemia and cyanosis may result from mucous plugging of the lower bronchial tree, atelectasis, and V/Q mismatch in infants with acute RSV infection. The hypoxemia is usually transient and not life-threatening. Hypoxemia is readily corrected with supplemental oxygen administration.

(Choice 5) Wheezing is a very common sign of RSV infection. It signifies inflammation of the bronchioles, but it is not life-threatening.

135. Question

1 points

Category: Pediatrics

A 2-month-old male infant is rushed to the emergency department by his baby sitter. She says that the baby became unresponsive and blue while feeding. The whole episode lasted 45-60 seconds, but the baby still appears pale. She denies any history of coughing or choking. On examination, the infant appears cyanotic and tachypneic. His blood pressure is 90/60 mm of Hg; his pulse rate is 155/min and regular. On auscultation, a harsh pansystolic murmur is noted at the left sternal border, along with a single S2. Hepatomegaly is noted. Pulse oximetry shows an oxygen saturation of 80%. Based on this history and physical examination, what is the most likely diagnosis?

1. ☐ Truncus arteriosus
2. ☐ Ventricular septal defect
3. ☒ Tetralogy of Fallot ✓
4. ☐ Patent ductus arteriosus
5. ☐ Transposition of great vessels

INCORRECT ✗

The correct answer is 3.

Tetralogy of Fallot is the most common cyanotic congenital disease in children less than 4 years of age. Patients often present with cyanotic spells. The classic tetrad includes:

1. Overriding aorta
2. Right ventricular hypertrophy
3. Subpulmonary stenosis (single S2)
4. Ventricular septal defect (VSD) (pansystolic murmur).

Because a large un-restrictive VSD is always present, the right ventricular pressure is the same as the left ventricular and aortic pressures. Pulmonary artery pressure and flow are inversely proportional to the degree of subpulmonary obstruction. The degree of cyanosis correlates precisely with the amount of obstruction.

(Choices 2 & 4) Patent ductus arteriosus and VSD are non-cyanotic congenital heart diseases.

(Choice 5) Transposition of the great vessels is a common cause of death within the 1st year of life. Children with this condition develop cyanosis and tachypnea within the first few days of life.

136. Question

1 points

Category: Pediatrics

A 6-month-old infant is taken to the emergency department because he had a seizure. Physical examination demonstrates premature closure of cranial sutures and markedly bowed legs. Laboratory studies demonstrate low serum phosphate levels, with normal vitamin D and parathyroid hormone levels. Urinalysis shows high phosphate levels, but no increased excretion of glucose, amino acids, or protein. The child's maternal grandfather had crippling bone disease, and his mother has mild bowing of the legs. Which of the following is most likely diagnosis?

1. ☐ Fanconi syndrome
2. ☒ Hypophosphatemic rickets ✓
3. ☐ Osteogenesis imperfecta
4. ☐ Osteomalacia
5. ☐ Paget disease of bone

INCORRECT ✖

The correct answer is 2.

This is hypophosphatemic rickets, also known as vitamin D-resistant rickets. This is a usually familial (often with X-linked dominant genetics) and rarely acquired abnormality of phosphate metabolism with impaired renal tubular resorption of phosphate accompanied by decreased intestinal absorption of calcium and phosphate. Vitamin D and parathyroid hormone levels are normal in these cases. The underlying biochemistry can be either impaired renal synthesis of 1, 25-dihydroxy-vitamin D₃, or impaired cellular response to this substance. Some patients may have only the hypophosphatemia (particularly heterozygous females), whereas others also show severe bone disease. Some patients, as the one in the question, present with craniostenosis (premature suture fusion with resulting abnormal head shape) and seizures before 1 year of age. Other patients present in childhood with bony abnormalities that can include bowed legs, other bone deformities, pseudo fractures, short stature, bone pain, and bony outgrowths at muscle attachments that may limit joint motion. This condition can now be treated with combined oral phosphate and 1, 25-dihydroxy-vitamin D₃

(Choice 1) can have hypophosphatemia as a component, but will show a generalized impairment of renal tubular absorption that also involves glucose, amino acids, and protein.

(Choice 3) causes brittle bones that fracture easily.

(Choice 4) is the term used for abnormalities of calcium and phosphate metabolism. It is similar to rickets and affects adults who have reached full stature.

(Choice 5) is a bony remodeling disease of adults.

137. Question

1 points

Category: Pediatrics

A 10-month-old girl is seen in the clinic for a routine checkup. She weighs 10 kg (22 lb) and is 72 cm tall. The mother reports that the infant takes solid foods poorly. Therefore, she is supplementing solid foods with whole cow milk because she can no longer produce breast milk. The mother notes that the infant has increasing lethargy and exhibits less activity compared with other children of the same age. The infant has had three upper respiratory tract infections and one episode of acute otitis media. She is otherwise sleeping well and has met developmental milestones appropriate for her age. On examination, she is found to have pallor of the skin and mucous membranes, good muscle tone, and normal neurologic function. Routine urinalysis is normal, and a peripheral blood smear shows microcytic and hypochromic red blood cells. Complete blood count shows the following:

Mean corpuscular volume (MCV): 77 fL

Mean corpuscular hemoglobin concentration (MCHC): 27 g/dL

Platelet count: 445,000/ μ L

Hematocrit: 30%

Red cell distribution width: 16.5

Which of the following is the most appropriate next step in management?

1. ☐ Antibiotic therapy
2. ☐ Bone marrow biopsy
3. ☒ Dietary advice and oral iron treatment
4. ☐ Red cell enzyme studies
5. ☐ Thyroid studies

INCORRECT ✖

The correct answer is 3.

Because the most likely clinical diagnosis at this time is iron deficiency anemia, it would be appropriate to offer dietary advice (iron-fortified cereal, meat, green vegetables, and egg yolk) and provide supplemental iron in the form of drops. A reticulocytosis typically occurs with a peak response in 5 to 7 days, followed by an increase in hemoglobin over the next 1 to 4 weeks. This treatment is both diagnostic and therapeutic.

(Choice 1) may also be used in hookworm infections to prevent further loss of blood and may be useful in the treatment of other concurrent infections, but it will have no effect in this child's nutritional iron deficiency anemia due to cow milk ingestion.

(Choice 2) is helpful in evaluating the presence of malignant cells and bone marrow failure, in the case of leukemia.

(Choice 4) are used to diagnose glucose 6-phosphate (G6PD) deficiency and pyruvate kinase as the cause of intrinsic hemolysis and subsequent anemia.

(Choice 5) can diagnose hypothyroidism as a cause of anemia. However, clinical and laboratory features of this child's anemia suggest nutritional iron deficiency.

138. Question

1 points

Category: Pediatrics

A 4-year-old Hispanic boy is brought to the office by his parents because he has difficulty in walking. The child started walking at the age of 17 months. He often falls during walking and has a tendency to run on his toes. He has relatives with a similar symptomatology and a maternal uncle who "had a premature death." Examination reveals weakened hip muscles, hypertrophied calves with wooden consistency, and positive Gower's sign. What is the best test to confirm this patient's diagnosis?

1. ☐ Creatine kinase levels
2. ☐ Electromyography
3. ☒ Muscle biopsy ✓
4. ☐ Nerve conduction studies
5. ☐ Serum aldolase levels

INCORRECT ✗

The correct answer is 3.

The patient most likely has Duchenne muscular dystrophy (DMD), which is the most common muscular dystrophy. His manifestations (i.e., pseudohypertrophy of the calves and positive Gower's sign) are characteristic for the disease. Furthermore, the similar symptomatology in the maternal uncle points out an X-linked recessive transmission. Muscle biopsy can confirm the diagnosis in this patient, although performing genetic studies are currently the most appropriate way of confirming the diagnosis in atypical cases.

(Choice 1) An elevated serum creatinine kinase level is probably the earliest manifestation of the disease. Increased levels can be seen in newborns. Similarly, aldolase levels are elevated; however, these levels can drop as more muscle gets replaced with fat and fibrosis.

These tests are typically used for screening purposes, rather than confirmatory.

(Choices 2 & 4) In DMD, EMG demonstrates a characteristic myopathic pattern with normal nerve conduction velocities; however, it is not used to confirm the diagnosis.

139. Question**1 points****Category: Pediatrics**

A 6-year-old male presents to the physician with eyelid swelling. His mother noted an insect bite on his left cheek yesterday, and states that this morning his left eyelid was completely swollen shut and painful. He has not had a fever, fatigue, headache, or rhinorrhea. On examination, he is afebrile. The left eyelid is erythematous, tender to palpation, and swollen shut. When trying to clinically distinguish between preseptal and orbital cellulitis, which of the following findings is most likely to be seen with preseptal cellulitis?

1. ☐ Decreased eye movements
2. ☐ Decreased visual acuity
3. ☐ Proptosis
4. ☒ Eyelid discoloration ✓
5. ☐ Fever

INCORRECT ✗**The correct answer is 4.**

In patients with cellulitis near the orbit and surrounding tissues, it is important to distinguish between preseptal cellulitis and orbital cellulitis. Preseptal cellulitis is an infection of the eyelid anterior to the orbital septum, whereas orbital cellulitis occurs when the infection is posterior to the orbital septum. Both types of cellulitis can be caused by local trauma (such as an insect bite or wound) or by extension from another source of infection including sinusitis or dental infections. Clinically, both preseptal and orbital cellulitis can present with tenderness, erythema, warmth, swelling, or eyelid discoloration but eyelid symptoms (e.g. discoloration) are more common in preseptal cellulitis. Orbital cellulitis can be distinguished from preseptal cellulitis by the presence of any of the following: decreased eye movements, decreased visual acuity, double vision, or proptosis. In addition, fever, malaise, and systemic symptoms are more common in orbital cellulitis than preseptal cellulitis.

When the differentiation of preseptal cellulitis and orbital cellulitis cannot be made clinically, a CT scan of the orbits is the imaging modality of choice to determine if the infection has spread posterior to the orbital septum. The mainstay of treatment for both preseptal and orbital cellulitis is antibiotics. While children with preseptal cellulitis may be managed as an outpatient if orbital cellulitis has been ruled out and they are otherwise well, orbital cellulitis requires inpatient treatment with intravenous antibiotics and close monitoring. In patients who fail to respond to antibiotics or have progressive visual loss, surgical decompression is indicated.

(Choice 1) Although pain with eye movements can be seen in patients with either preseptal cellulitis or orbital cellulitis, decreased eye movements are usually only seen in patients with orbital cellulitis.

(Choices 2 & 3) Decreased visual acuity and proptosis are both findings seen in patients with orbital cellulitis, not preseptal cellulitis.

(Choice 5) Although fever can occur in both conditions, it is much more common in patients with orbital cellulitis.

140. Question

1 points

Category: Pediatrics

A premature neonate develops respiratory distress syndrome several hours after birth. The infant is placed on a respirator and given other appropriate care. However, when the infant reaches a corrected gestational age of 36 weeks, he does not tolerate weaning from the ventilator. A chest x-ray film demonstrates alternating areas of hyperaeration and pulmonary scarring, resulting in parenchymal streaks and hyper expanded areas. Which of the following is the most likely diagnosis?

1. ☐ Apnea of prematurity
2. ☒ Bronchopulmonary dysplasia 
3. ☐ Cystic fibrosis
4. ☐ Persistent pulmonary hypertension of the newborn
5. ☐ Transient tachypnea of the newborn

INCORRECT 

The correct answer is 2.

This infant has bronchopulmonary dysplasia, which is an important chronic lung disorder that can complicate respiratory distress syndrome of the newborn. Damage to the lungs by mechanical ventilation produces alternating areas of emphysema and scarred lungs. The smooth muscles of bronchioles and arterioles may also be hypertrophied. The modern trend is to try to prevent this complication by using the smallest amount of mechanical ventilation that still adequately aerates the infant's lungs. Management of significant degrees of bronchopulmonary dysplasia is problematic, and therapeutic regimens vary from neonatal unit to neonatal unit.

(Choice 1) causes transient interruptions in breathing.

(Choice 3) The lung problems of cystic fibrosis are usually not present at birth, but develop after the child has repeated bouts of pneumonia.

(Choice 4) Persistent pulmonary hypertension of the newborn is due to a persistence of fetal type circulation (typically due to either a patent foramen ovale or a persistent patent ductus arteriosus), which clinically causes severe hypoxemia.

(Choice 5) causes 2 or 3 days of difficult breathing in neonates who are slow to reabsorb amniotic fluid trapped in the lungs.

141. Question

1 points

Category: Pediatrics

A 7-year-old boy is rushed to the emergency department after falling on his outstretched hand. He immediately complained of right arm pain after the accident, and he currently cannot move his arm due to the pain. He is crying and holding his right arm in flexion. There is ecchymosis just above his elbow. He cries out in pain when his arm is moved. An x-ray reveals a supracondylar fracture. What secondary injury is most commonly associated with this patient's fracture?

1. ☐ Median nerve injury
2. ☐ Axillary nerve injury
3. ☐ Ulnar artery injury
4. ☒ Brachial artery injury ✓
5. ☐ Brachial plexus injury

INCORRECT ✗

The correct answer is 4.

Supracondylar fractures are the most common fractures in the pediatric population. It is rarely seen in adults, and the majority result from trauma. The most common complication is entrapment of the brachial artery, resulting in loss of the radial artery pulse; therefore, all patients with a supracondylar fracture must have their radial pulse checked. After reduction of the fracture, the radial pulse must be reassessed.

(Choice 1) The median nerve is spared in a supracondylar fracture. Once the fracture is reduced, the edema subsides and the paresthesias in the arm subside.

(Choice 2) The axillary artery is injured during proximal humerus fractures, but not with supracondylar fractures.

(Choice 3) The ulnar artery is not directly injured with supracondylar fractures. If the brachial artery is compromised with the fracture, the ulnar pulse will disappear.

(Choice 5) Brachial plexus injury is not common with supracondylar fractures.

142. Question

1 points

Category: Pediatrics

A 16-year-old College student presents to the physician's office because of a generalized malaise, sore throat and fever for the past 2 to 3 days. She is given amoxicillin prescription. Twenty-four hours later, she develops a polymorphous rash over her entire body. She has taken amoxicillin in the past for sinus disease and never had any problem. What is the most likely organism that is responsible for her symptoms?

1. ☐ Group B streptococcus
2. ☐ Epstein-Barr virus
3. ☒ Coxsackie virus ✓
4. ☐ Escherichia coli
5. ☐ Nocardia

INCORRECT ✗

The correct answer is 3.

Infectious mononucleosis is an infection caused by the Epstein-Barr virus, and is sometimes detected only when the patient develops a characteristic polymorphous rash after taking ampicillin or amoxicillin for an apparent upper respiratory tract infection. A prodrome of malaise, fever and adenopathy can usually be extracted from the history. The agent is spread via the saliva, and the condition usually resolves after two to three weeks.

(Choice 1) Group A β -hemolytic streptococci (not Group B) is responsible for 20-25% cases of pharyngitis in children. Group B streptococcus infections in the adult are serious. These are common in diabetics and those with peripheral vascular disease.

Complications include endocarditis, arthritis, pneumonia, empyema and meningitis. It does not present with a rash after taking amoxicillin.

(Choice 3) Coxsackie virus generally produces fever, headache and stiff neck (signs of aseptic meningitis). It can cause myocarditis, hand-foot-and-mouth disease, and herpangina (vesicles on the hard palate). It generally does not produce a rash after taking an antibiotic.

(Choice 4) Escherichia coli generally causes urinary tract infections. It can also cause infections anywhere in the abdominal cavity, and is particularly prominent in infections that involve ischemic extremities, surgical wounds and leukemia patients. It does not produce a rash.

(Choice 5) Nocardiosis usually occurs in patients who are immunocompromised and taking steroids. It presents as pneumonia with cavitary lesions. It can also present with multiple abscesses and draining sinuses. It is a partially acid-fast organism, and is sensitive to sulfonamides. It does not produce a rash.

143. Question

1 points

Category: Pediatrics

A term neonate is noted at birth to have multiple structural defects, many of which are midline in location. He has a bilateral cleft lip and palate, very small eyes with fissures of the iris, shallow supraorbital ridges, and slanted palpebral fissures. His ears are malformed and low set. Both hands have six fingers and a single simian crease, and his feet are shaped like the runner of a rocking chair. There is no family history of congenital abnormalities. At 1 month of age, he dies from heart failure. Which of the following neurologic abnormalities was likely to have been present in this infant?

1. ☐ Anencephaly
2. ☐ Encephalocele
3. ☐ Hydranencephaly
4. ☒ Holoprosencephaly
5. ☐ Porencephaly

INCORRECT ❌

The correct answer is 4.

This is trisomy 13, also known as Patau syndrome. The presentation illustrated is typical. These infants usually die before 1 year of age from multiple congenital anomalies, particularly severe congenital heart defects. Patau syndrome is associated with holoprosencephaly due to a failure of forebrain development and consequent failure to form paired cerebral hemispheres. It is associated with midface developmental abnormalities. Other features include hyperconvex, narrow fingernails, scalp defects, abnormal genitalia, apneic spells, holoprosencephaly, and severe mental retardation.

(Choice 1) is a complete failure of higher brain structures to develop. It is accompanied by a grossly deformed head with virtually no cranial vault.

(Choice 2) is protrusion of brain and meninges due to a developmental defect in the skull.

(Choice 3) is an extreme form of porencephaly in which the cerebral hemispheres are destroyed. Unlike in anencephaly, the external head forms normally.

(Choice 5) is a cyst or cavity in the brain that communicates with the ventricles; it may occur as a developmental anomaly or secondary to inflammatory disease or a vascular accident.

144. Question

1 points

Category: Pediatrics

A mother brings her 9-month-old daughter to the pediatrician with complaints of a rash. The mother states that the infant had a high fever (temperature up to 40.0 C [104.0F]) for 3 days prior to developing the rash, but is now afebrile. The mother also says that the infant has had a runny nose and a slight cough for the past 3 days. On examination, there is a fine macular rash on the infant's trunk and neck. The examination is otherwise within normal limits, and the infant is playful and smiling. Which of the following is the most likely diagnosis?

1. ☐ Erythema infectiosum
2. ☒ Roseola ✓
3. ☐ Rubella
4. ☐ Rubella
5. ☐ Varicella

INCORRECT ✗

The correct answer is 2.

Roseola is a virus that causes an exanthem (exanthem subitum). It typically affects children between 6 months and 3 years of age. Most cases manifest with a fever (temperature often up to 40-40.5C) that lasts for 2-3 days; there is often mild erythema of the pharynx and congestion. The discrete, macular rash typically erupts on the trunk with an abrupt end to the fever.

(Choice 1) Erythema infectiosum, also known as Fifth disease, is caused by parvovirus B 19. This virus usually affects children aged 5-14 years. There is usually a nonspecific febrile illness with malaise and myalgias lasting 2-3 days. The rash of erythema infectiosum is characterized by a red rash on the cheeks ("slapped" cheeks) and a erythematous maculopapular rash on the extremities and trunk described as a "lacy" reticular pattern.

(Choice 3) Rubella, also known as German measles, is characterized by a low-grade fever (temperature usually less than 39 C), erythematous maculopapular rash, and lymphadenopathy.

(Choice 4) Rubella, or measles, is characterized by 3-5 days of increasing fever, coryza, cough, and conjunctivitis. Koplik spots (white lesions on a red mucosa) appear in the mouth. There is a discrete, erythematous, maculopapular rash that begins on the face and moves downward, covering the trunk and upper and lower extremities.

(Choice 5) Varicella, or chickenpox, begins with a period of malaise and low-grade fever. The lesions begin as macules and progress to fluid filled vesicles that then crust; the crust eventually falls off. The fever is usually gone by the end of the first week of the rash.

145. Question**1 points****Category: Pediatrics**

An infant has had repeated pneumonias and middle ear infections that began at about 5 months of age. At 1 year of age, serum electrophoresis demonstrated hypogammaglobulinemia. T cell function was normal. By 2 years of age, the child's infection rate has decreased, and repeat serum electrophoresis is normal. Which of the following immunoglobulins was likely decreased in this child during the period of increased susceptibility to infection?

1. ☐ IgA
2. ☐ IgD
3. ☐ IgE
4. ☒ IgG
5. ☐ IgM

INCORRECT ❌**The correct answer is 4.**

The condition is called transient hypogammaglobulinemia of infancy. It is due to exhaustion of maternally supplied IgG (the only antibody to cross the placenta in significant amounts) before the infant has begun to produce significant amounts of his own antibodies. Affected infants typically go through several months to years of being very vulnerable to infection, and then improve as their immune systems mature and they are able to produce more IgG. The condition does not present in the neonatal period because maternal IgG can cross the placenta during the third trimester of intrauterine life. The maternally supplied IgG is usually exhausted by about the 4th to 6th month of life.

(Choice 1) is found in secretions and in serum but does not cross the placenta.

(Choice 2) is a transiently produced antibody during B cell development that is present in trace amounts in serum.

(Choice 3) is the antibody associated with allergic reactions; it does not cross the placenta.

(Choice 5) is a large antibody found in serum that does not cross the placenta.

145. Question

1 points

Category: Pediatrics

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146. Question

1 points

Category: Pediatrics

A 4-month-old infant is evaluated by a dermatologist because of thick, erythematous skin with fine scaling, principally involving his face. The mother reports that the infant is "always scratching his face." An older brother and a maternal uncle had a similar condition. Screening hematologic studies show the following:

Erythrocyte count: 5.1 million/mm³

Leukocyte count: 12,000/mm³

Segmented neutrophils: 80%

Bands: 5%

Eosinophils: 3%

Basophils: 1%

Lymphocytes: 5%

Monocytes: 6%

Platelet count: 35,000/mm³, with the comment that the platelets are smaller than normal

Serum immunoglobulin studies demonstrate the following:

IgA: 120 mg/dL,

IgE: 2300 IU/mL

IgG: 900 mg/dL

IgM: 15 mg/dL

Patients with this condition have a significantly increased incidence of which of the following?

1. ☐ Basal cell carcinoma
2. ☐ Hodgkin lymphoma
3. ☐ Melanoma
4. ☒ Non-Hodgkin lymphoma ✓
5. ☐ Squamous cell carcinoma of the skin

INCORRECT ✗

The correct answer is 4.

The disease is Wiskott-Aldrich syndrome. This X-linked condition is characterized by thrombocytopenia (with characteristically small platelets) and lymphopenia with depressed cellular immunity. The lymphocytes have a reduced level of the adhesion molecule sialophorin (CD43) on their surfaces, which forms the basis of a helpful diagnostic test in this condition. Atopic eczema, as seen in this child, is very common and may be the presenting complaint in these patients. It is typically accompanied by high serum IgE, often with low serum IgM. If the thrombocytopenia is very severe, splenectomy may be helpful in controlling the bleeding tendency. For reasons that are not yet biochemically clear, but have to do with their already abnormal lymphoid cells, these patients are very prone to develop non-Hodgkin lymphomas. They do not have a significantly increased tendency to develop skin malignancies, such as melanomas, squamous cell carcinomas, and basal cell carcinomas.

(Choice 1) Associate basal cell carcinoma with xeroderma pigmentosa and basal cell nevus syndrome.

(Choice 2) Hodgkin disease does not have any noteworthy associations with genetic diseases.

(Choice 3) Associate melanoma with xeroderma pigmentosa and very large congenital nevi.

(Choice 5) Associate squamous cell carcinoma of the skin with xeroderma pigmentosa.

147. Question**1 points****Category: Pediatrics**

A 16-year-old girl is brought to the office due to abdominal pain and bloody diarrhea for the past two days. Her immunizations are up to date. Her past medical history is unremarkable. On examination, she appears lethargic and weak. Her skin turgor is poor, face and extremities are swollen, and skin appears yellow. Abdominal examination shows moderate non-localized, abdominal pain. Laboratory studies show:

Hb: 64 g/dL**Hct:** 26.9%**Platelets:** 45,000/mm³**Creatinine:** 2.3 mg/dL**Total bilirubin:** 5.0 mg/dL

What is the most common complication of the patient's condition?

1. ☒ Renal damage ✓
2. ☐ Liver damage
3. ☐ Bone marrow failure
4. ☐ Pneumonia
5. ☐ Bowel obstruction

INCORRECT ✗

The correct answer is 1.

Hemolytic-uremic syndrome (HUS) is caused by a toxin released from *Escherichia coli*. The toxin enters the systemic circulation and injures the endothelial cells in the kidney. When there is progressive elevation of BUN and creatinine, the chances for improvement of the kidneys' function is poor. When the kidneys are damaged, the mortality is 5-10%.

(Choice 2) The liver is not directly damaged by the toxin in HUS. The jaundice seen in HUS is due to hemolysis, and not liver injury.

(Choice 3) The bone marrow is relatively spared from the toxin. The thrombocytopenia and anemia are due to intravascular destruction of the RBCs and platelets.

(Choice 4) Pneumonia is not a clinical feature of HUS.

(Choice 5) Bowel obstruction does not occur with HUS. The usual presentation is abdominal cramping and bloody diarrhea.

148. Question**1 points****Category: Pediatrics**

A 2-year-old boy who emigrated from Eastern Europe 1 year ago is brought to the physician because of fever, cough, and night sweats for 3 weeks. The child's grandmother, who lives with him, has similar symptoms. The child's temperature is 39.2 C (102.6 F), blood pressure is 110/65 mm Hg, pulse is 90/min, and respirations are 28/min. A Mantoux test is reactive, and a chest x-ray film shows a right middle lobe infiltrate and hilar lymphadenopathy. Which of the following is the most appropriate next step in diagnosis?

1. ☐ Cervical lymph node biopsy
2. ☒ Gastric aspiration ✓
3. ☐ Pleurocentesis
4. ☐ Sputum induction

INCORRECT ✗**The correct answer is 2.**

This child has primary pulmonary tuberculosis. The infection was most likely acquired from his grandmother, who is sick with similar symptoms. The initial pneumonic area may be anywhere in the lung, especially in the middle and lower lobes. This age group of patients rarely has a positive Mantoux test in the absence of recent infection. Early morning gastric aspiration is the best method for identifying acid-fast bacilli, which are swallowed during the night. Gastric aspiration is optimal in the early morning before the gastric acid destroys the bacilli.

(Choice 1) This patient does not have miliary tuberculosis, and so a cervical lymph node biopsy would not be of diagnostic value.

(Choice 3) This patient has does not have evidence of a pleural effusion; therefore, pleurocentesis is not indicated.

(Choice 4) It would be difficult for a 2-year-old to comply with sputum induction. In addition, sputum induction may be negative in more than 50% of cases of pulmonary tuberculosis.

149. Question

1 points

Category: Pediatrics

A 3-week-old infant is being evaluated for hematochezia. His mother states that the infant passed stools that contain both blood and mucus. There were no complications during her pregnancy, and the infant has been otherwise healthy. On physical examination, his temperature is 37.1 C (98.9 F), pulse is 110/min, and respirations are 18/min. He appears well, and his fontanelle is flat and level. Abdominal examination reveals normal active bowel sounds; his abdomen is nontender to palpation and there is no mass. His diaper contains stool that has bright red blood on it with mucus. Which of the following is the most likely explanation of his hematochezia?

1. ☒ Food allergy-induced colitis ✓
2. ☐ Meckel diverticulum
3. ☐ Necrotizing enterocolitis
4. ☐ Rectal fissure
5. ☐ Ulcerative colitis

INCORRECT ✗

The correct answer is 1.

Food allergy-induced colitis is a clinical syndrome of enterocolitis usually presenting between 1 week and 3 months of age. The dietary proteins implicated are generally cow's milk or soy proteins. Although diarrhea and emesis are the most prominent symptoms, rectal bleeding, protein-losing enteropathy with edema, and dehydration with metabolic acidosis are also prominent. The diarrhea is relatively acute and progressive to the point of dehydration if the etiologic protein is not eliminated. The stools contain mucus, polymorphonuclear leukocytes, and eosinophils. Rectal bleeding progresses from occult to increasingly gross bleeding and anemia. Whereas the diarrhea is most consistent with an inflammatory colitis, stools may also contain reducing sugars, suggesting a small intestinal component as well. Approximately one third of infants with severe diarrhea will develop metabolic acidosis. Hypotension may complicate the presentation in younger infants but is more commonly seen following protein challenge, especially with soy.

Irritability, emesis, and abdominal distention are also prominent with acute manifestations. When a more indolent clinical course is encountered, the infant will manifest failure to thrive, edema from enteric protein loss, and anemia. Food refusal, a feature of eosinophilic esophagitis and enteropathy, is also common. The diagnosis is confirmed when symptoms resolve within 72-96 hours after the elimination of the offending protein from the diet, and subsequent oral challenge provokes both symptoms and a measurable inflammatory response. More than 90% of the infants will respond to initiation of an extensively hydrolyzed casein-based formula. From 15% to 50% of infants sensitive to cow's milk will also be sensitive to soy-based formula.

(Choice 2) is the most common congenital malformation of the gastrointestinal tract, occurring in about 1.5% of the general population, most of whom experience no symptoms. The mucosa is often gastric, and the acid secreted may cause ulceration and bleeding, which is usually painless.

(Choice 3) affects premature infants and usually occurs as a complication of respiratory distress syndrome or patent ductus arteriosus. It manifests as abdominal distention, tenderness, bilious vomiting, and occult or frank blood in stool.

(Choice 4) is one of the most common causes of rectal bleeding in young children. There is usually a history of constipation, with passage of large and hard stools.

(Choice 5) is an inflammatory bowel disease primarily affecting the colon. It causes abdominal pain, hematochezia, diarrhea, weight loss, and fever.

150. Question**1 points****Category: Pediatrics**

A 5-year-old child undergoes a school entrance physical examination. The pediatrician notices grey-brown pigmentation on the skin of his forehead, hands, and pretibial regions. Subconjunctival areas near the corneoscleral junction show wedge-shaped, yellow-brown discoloration (pinguecula). Enlargement of both the spleen and the liver are noted on abdominal examination. Needle biopsy of the spleen demonstrates the presence of unusually large (20- to 100-mm diameter) reticuloendothelial histiocytes with a "crumpled-silk" appearance. Bone marrow biopsy demonstrates the presence of the same type of cells. Which of the following is the most likely diagnosis?

1. ☐ Abetalipoproteinemia
2. ☐ Fabry disease
3. ☒ Gaucher disease ✓
4. ☐ Niemann-Pick disease
5. ☐ Tangier disease

INCORRECT ✗**The correct answer is 3.**

This is Gaucher disease, which is a familial autosomal recessive disorder of lipid metabolism. The disease is caused by a lack of glucocerebrosidase activity, which normally hydrolyzes glucocerebroside to glucose and ceramide, and occurs in three major clinical forms. All three types are characterized by prominent splenomegaly and accumulation of abnormal glucocerebrosides in reticuloendothelial cells in many organs (spleen, liver, bone marrow, and brain in more severe cases), producing the pathognomonic "crumpled-silk" histiocytes. This case is an example of the type 1, or the adult chronic non-neuronopathic, form. Type 1 is the most common form and actually presents frequently in childhood, although the most serious manifestations are often deferred to adulthood. Type 1 has an increased frequency in Ashkenazi Jews and manifests with hypersplenism, splenomegaly, and bone lesions. The pingueculae and brown skin pigmentation noted in the question stem may be helpful clues on physical examination. Type 2, the acute infantile neuronopathic form, causes splenomegaly, severe neurologic abnormalities, and death in early childhood. Type 3 is a juvenile form and has features intermediate to types 1 and 2.

(Choice 1) is characterized by absent β -lipoproteins, steatorrhea, acanthocytes, retinitis pigmentosa, ataxia, and mental abnormalities.

(Choice 2) is characterized by angio-keratomas, corneal opacities, burning extremity pain, and involvement of kidneys, heart, and brain.

(Choice 4) may clinically resemble Gaucher disease, but is characterized by "sea-blue" rather than "crumpled-silk" histiocytes.

(Choice 5) may resemble Gaucher disease, with hepatosplenomegaly and neurologic disease, but a helpful distinctive feature is orange-yellow tonsillar hyperplasia.

151. Question

1 points

Category: Pediatrics

A 2-week-old boy in the neonatal intensive care unit had a birth weight of 1200 g. Ultrasound of the head reveals grade II intraventricular hemorrhage and periventricular leukomalacia. An ophthalmologic examination reveals retinopathy of prematurity of both eyes. In addition, a hearing screen, demonstrates bilateral hearing deficits. Which of the following is the most important determinant of this child's neurodevelopmental outcome?

1. ☒ Length of gestation
2. ☐ Maternal education
3. ☐ Outcome of the mother's previous pregnancies
4. ☐ Quality of prenatal care
5. ☐ Socioeconomic status of the family

INCORRECT ❌

The correct answer is 1.

The length of gestation is the single most important determinant of the neuro-developmental outcome of any very low-birthweight (<1500 g) infant. Infants born before 23 weeks' gestation have an 85% perinatal mortality rate. Mortality decreases to about 25% at 25 weeks.

(Choice 4) also plays a significant role in the neurodevelopmental outcome of the infant. Improved obstetric care has reduced the rate of perinatal asphyxia. Antenatal administration of corticosteroids to mothers who might have preterm labor has decreased the incidence and severity of respiratory distress syndrome (hyaline membrane disease). However, the impact of prenatal care impact is not as great as that of the length of gestation.

(Choices 2 & 5) Maternal education and socioeconomic status are also found to have some impact on the long-term neurodevelopmental outcome of infants. Limited financial and educational resources, as well as suboptimal home environment, may have contributed to this finding.

(Choice 3) has not been shown to have significant impact on the long-term neurodevelopmental outcome of a very low-birthweight infant. Nonetheless, a previous preterm delivery is a risk factor for a future preterm pregnancy.

151. Question

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INCORRECT ✗

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152. Question

1 points

Category: Pediatrics

A 3-year-old child presents to the emergency department via ambulance with seizures followed by coma. The patient is initially stabilized, and diagnostic studies are obtained. The findings of the cerebrospinal fluid (CSF) include a total white blood cell (WBC) count of 250 cells/ μ L (normal, 0-5 cells/ μ L) with a predominance of lymphocytes and monocytes. The CSF glucose is 20 mg/dL (normal, 40-70 mg/dL), the total protein concentration is 50 mg/dL (normal, 15-45 mg/dL), and the chloride concentration is 105 mEq/L (normal, 118-132 mEq/L). Results of a Gram stain of spun-down CSF sediment are negative. A computed tomography (CT) scan with contrast shows enhancement at the base of the brain. Which of the following is the most likely diagnosis?

1. ☐ Eastern equine encephalitis
2. ☐ Subacute sclerosing panencephalitis
3. ☒ Tuberculosis meningitis ✓
4. ☐ Cryptococcal meningitis
5. ☐ Meningococcal meningitis

INCORRECT ✗

The correct answer is 3.

Tuberculosis meningitis usually presents in recently infected individuals and is usually considered a disease of infants and children. The symptoms develop slowly in three stages. Stage 1 is a prodromal stage with nonspecific symptomatology. Seizures are common in stage 2; coma occurs in stage 3. A high index of suspicion is necessary for rapid diagnosis. A computed tomography (CT) scan of the head may show periventricular lucencies, edema, infarctions, and hydrocephalus. Cerebrospinal fluid (CSF) findings include an elevation in white blood cells (WBCs), usually from 250 to 500 cells/L, predominantly lymphocytes. The CSF glucose is less than 40 mg/dL, or less than half the serum glucose found simultaneously. Protein concentration is normal or slightly high. Gram's stain is negative because the bacilli are acid-fast. CSF chloride is low.

(Choice 4) is associated with elevated protein, hypoglycorrhachia, and mononuclear pleocytosis (rarely >300 cells/ mm^3).

(Choice 2) Children with subacute sclerosing panencephalitis have CSF with normal white cell content (may show some plasma cells), normal or slightly elevated protein, and greatly increased globulin.

(Choice 1) occurs more commonly in young infants, causing serious complications and fatalities.

(Choice 5) The spectrum and definitive diagnosis of meningococcal meningitis are made by the isolation of the organism from the CSF.

153. Question

1 points

Category: Pediatrics

A 3-month-old male infant is brought to the emergency department by his mother because of difficulty in breathing. His mother was admitted in the hospital ten days ago due to a urinary tract infection, and he was cared for by his grandmother during that period. His mother had just been discharged from the hospital yesterday, and noticed that he was constipated and having difficulty with breastfeeding. On examination, he is afebrile. His pulse rate is 110/min, respirations are 36/min with shallow breathing efforts, and blood pressure is 90/50mm Hg. His weight is at the 35th percentile. Examination shows ptosis, dilated pupils with sluggish reaction to light, diminished deep tendon reflexes and decreased muscle tone. What is the most likely mechanism of his illness?

1. ☐ Bacterial infection of the meninges
2. ☐ Autoimmune disease against acetylcholine receptors
3. ☐ Clostridium difficile toxin in the intestinal tract
4. ☒ Clostridium botulinum in the intestinal tract
5. ☐ Clostridium botulinum toxin intake

INCORRECT ❌

The correct answer is 4.

This is a typical case of infant botulism. *C. botulinum* is the most common etiologic agent and is usually transmitted through food, especially honey, which is the carrier in 1/3 of cases. In the infantile form of botulism, the causative organism gains entry through the food and produces the toxin in the intestinal tract, which subsequently triggers the symptoms. It is a protease that blocks acetylcholine release. Infants typically present between two weeks and nine months with constipation and poor feeding. This is followed by progressive hypotonia, weakness, loss of deep tendon reflexes, cranial nerve abnormalities (impaired gag reflex), and respiratory difficulties. Signs of autonomic dysfunction such as hypotension and neurogenic bladder can occur early in the course of the disease.

(Choice 1) Patients with meningitis are typically febrile.

(Choice 2) Autoantibodies against acetylcholine receptors is a feature of myasthenia gravis, which presents with early fatigability of the skeletal muscles.

(Choice 3) *C. difficile* toxin is responsible for pseudomembranous colitis. Patients usually have diarrhea (not constipation), and most cases are associated with prior antibiotic use.

(Choice 5) The infantile form of botulism is different from the adult form. In the latter, the toxin ingested per se produces the symptoms.

154. Question

1 points

Category: Pediatrics

A frantic mother telephones the pediatric office. She reports that her 10-year-old boy accidentally splashed Drano (a strongly corrosive, alkaline drain cleaner) on his face, and he is screaming in pain complaining that his right eye hurts terribly. Which of the following is the best advice to give to the mother?

1. ☐ Apply antibiotic ointment to the eye and make an appointment with an ophthalmologist
2. ☐ Bring the boy to the hospital right away
3. ☐ Pry the eye open and drip vinegar over it until the pain goes away
4. ☐ Pry the eye open and swipe it clean with a tissue before bringing the boy in for further evaluation
5. ☒ Pry the eye open, hold it under running cold water for about 30 minutes, and then bring the boy to the hospital ✓

INCORRECT ✗

The correct answer is 5.

Alkaline burns are extremely destructive, and the process of destruction continues as long as the agent is in contact with the tissues. Immediate removal is essential, and the best way to do it is with massive irrigation. In the emergency department, sterile saline would be used; at home, tap water will do.

(Choice 1) Antibiotics do not help, and a leisurely approach will guarantee loss of the eye.

(Choice 2) Rushing to the hospital might seem like the thing to do for all kinds of emergencies, but in this case immediate first aid is more important. If the corrosive substance remains in the eye during the trip to the emergency department, the eye will be destroyed by the time the patient arrives.

(Choice 3) "Playing chemist" is never acceptable because of the possibility of causing an exothermic reaction.

(Choice 4) Swiping with a tissue would be better than nothing, but not by much. Swiping has a role, but not as the first thing to do. After massive irrigation, little particulate matter may remain, which may then have to be mechanically wiped away.

155. Question

1 points

Category: Pediatrics

A 10-month-old boy develops an upper respiratory tract infection 2 days before presentation. On the day of presentation, he has a generalized tonic-clonic seizure lasting 30 seconds. His temperature is 40.0 C (104.0 F), blood pressure is 90/60 mm Hg, and respirations are 22/min. He is alert and smiling. He has rhinorrhea, and his neck is supple. He has bruises below his knees. Which of the following is the most likely diagnosis?

1. ☐ Child abuse
2. ☐ Idiopathic epilepsy
3. ☐ Infantile spasms
4. ☐ Meningitis
5. ☒ Simple febrile seizure ✓

INCORRECT ✗

The correct answer is 5.

The most frequent cause of a short generalized tonic-clonic seizure is a simple febrile seizure. The patient must be treated with antipyretics, but prognosis is excellent. Febrile seizures are not considered to be epileptic since they are provoked events. This seizure type is seen typically in children aged 6 months to 3 years, but may occur until 6 years.

(Choice 1) Given the high proportion of child abuse cases seen in emergency departments, child abuse should always be on the differential. However, this patient's history suggests other causes.

(Choice 2) is also called the benign focal epilepsy of childhood. The age of onset is 4-10 years, and the disorder is not associated with underlying cerebral abnormalities.

(Choice 3) Infantile spasms are massive myoclonic seizures with either forceful flexion or extension of the trunk. The spasms occur in clusters, and the child may cry or be extremely irritable during these periods.

(Choice 4) should be considered, but a lumbar puncture is not essential in this infant, who is alert and smiling and showing no meningeal signs on examination.

156. Question

1 points

Category: Pediatrics

A 3-year-old girl is brought to the pediatrician with complaints of abdominal pain and fever. Her mother states that the fever started 2 days ago, with the highest temperature being 39.0 C (102.2 F). She has had no vomiting or diarrhea. The mother states that her daughter has been complaining of pain on urination. On examination, she is tender in her lower abdomen, and there is some right-sided costovertebral angle tenderness. A urinalysis confirms the suspicion of a urinary tract infection. Which of the following would be the most appropriate diagnostic procedure?

1. ☐ Cystoscopy
2. ☐ Dimercaptosuccinic acid (DMSA) scan in 1 to 2 months
3. ☐ Intravenous pyelogram
4. ☐ Voiding cystourethrogram (VCUG) now
5. ☒ VCUG in 1 to 2 months

INCORRECT ❌

The correct answer is 5.

A voiding cystourethrogram (VCUG) is the appropriate test to run to evaluate the lower urinary tract and the presence of any reflux of urine into the kidney. It is appropriate to first wait until the acute infection is over, since there may be some degree of reflux present during an infection. The usual procedure is to wait 1-2 months after treatment to perform the VCUG to evaluate for reflux.

(Choice 1) is not indicated in the evaluation of urinary tract infection.

(Choice 2) Dimercaptosuccinic acid (DMSA) scan is a radionuclide renal imaging scan. It might be an appropriate test to diagnosis acute pyelonephritis, but it is not indicated as a first test in evaluating reflux in a urinary tract infection 2 months after the infection. It is the most accurate test to diagnose acute pyelonephritis.

(Choice 3) Intravenous pyelogram is rarely used in the pediatric population to evaluate for a urinary tract infection.

(Choice 4) Performing a VCUG now would not be appropriate since there might be some degree of reflux present during an acute infection.

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(Choice 3) Intravenous pyelogram is rarely used in the pediatric population to evaluate for a urinary tract infection.

(Choice 4) Performing a VCUG now would not be appropriate since there might be some degree of reflux present during an acute infection.

157. Question

1 points

Category: Pediatrics

A 4-day-old girl is brought to the pediatrician because she has a blood-tinged discharge from her vagina. The patient was born at 38 weeks' gestational age to a 22-year-old primigravida. The infant weighed 7 lb 6 oz at birth and had no complications at birth. The infant was discharged home with her mother 24 hours after delivery. The mother reports that, since that time, the child has been afebrile and is feeding and sleeping well. At this point, the physician should do which of the following?

- ☐ Draw blood for a hematocrit determination.
- ☐ Notify the Child Protective Services.
- ☒ Reassure the mother that this is normal. ✓
- ☐ Call the endocrinologist.
- ☐ Obtain a urinalysis.

INCORRECT ✗

The correct answer is 3.

Vaginal bleeding, or pseudo-menses, may occur in a newborn female. In utero, the vaginal epithelium is stimulated by circulating maternal estrogens, which diffuse across the placenta. After birth, these hormone levels rapidly fall, which results in endometrial sloughing. There may be a thick mucoid discharge from the newborn's vagina, which is sometimes bloody. The discharge will usually resolve within 10 days after birth.

(Choices 2 & 3) No testing or treatment is necessary (choices 1 and 5), only reassurance. Because the vaginal bleeding is not related to abuse, the Child Protective Services need not be informed.

(Choice 4) It is not necessary to call an endocrinologist because this is not an endocrinopathy.

158. Question

1 points

Category: Pediatrics

A 3-year-old child is evaluated for possible causes of her mental retardation. Physical examination demonstrates elfin facies. The child has a history of stenosis of the aortic valve and transient hypercalcemia in infancy. Genetic micro deletion studies are performed. The lesion most likely involves which of the following genetic loci?

1. ☒ 7q11.23 ✓
2. ☐ 20p12
3. ☐ 22q11.21
4. ☐ Maternal chromosome at 15q11
5. ☐ Paternal chromosome at 15q11

INCORRECT ✗

The correct answer is 1.

Mental retardation associated with abnormal facial features may be related to abnormal genetics. As our skill with DNA analysis has improved, we have identified a number of micro deletion syndromes involving contiguous genes on a single chromosome that are associated with mental retardation and often distinctive clinical presentations. The case illustrated is William's syndrome, which involves 7q11.23.

(Choice 2) is associated with Alagille syndrome, characterized by lack of bile ducts, cardiac abnormalities, butterfly vertebrae, and eye abnormalities.

(Choice 3) is associated with DiGeorge syndrome, which is characterized by thymus and parathyroid abnormalities, heart defects, cleft palate, and mental retardation.

(Choice 4) Involvement of the maternal chromosome 15 at 15q11 is associated with Angelman syndrome, which is characterized by seizures, puppet-like movements, outbursts of laughter, and severe mental retardation.

(Choice 5) Involvement of the paternal chromosome 15 at 15q11 is associated with Prader-Willi syndrome, which is characterized by obesity, small hands and feet, and mental retardation.

159. Question

1 points

Category: Pediatrics

A 3-year-old child with history of chronic headache is brought to the pediatrician by her mother. The mother states that the child had been in her usual state of health until 2 weeks ago, when she started to complain of a continuous headache. The mother states that she has given the patient over-the-counter medication for the headache but nothing seems to make the headache better. In fact, the mother thinks that perhaps the headache seems to be worse. On review of symptoms, the mother states that the patient has had to "pee" with increasing frequency. The physician notes that the child is short, the patient's height is below the 5th percentile for her age. Although not very cooperative with the eye examination, the patient appears to have a bi-temporal hemianopia. The physician orders a computed tomography (CT) scan of the head that shows a large suprasellar mass. (Figures A and B below.) Based on the history and physical examination, as well as on the result of the CT scan of the head, this patient most likely has which of the following?

1. ☐ An astrocytoma of the third ventricle
2. ☐ A pinealoma
3. ☒ A craniopharyngioma
4. ☐ Chromophobic adenoma
5. ☐ Hand-Schüller-Christian disease

INCORRECT ❌

The correct answer is 3.

Craniopharyngioma is the most common supratentorial tumor of children. About 90% of these tumors show calcifications on plain skull films or on computed tomography (CT) scanning. Many cases are originally discovered during an endocrine workup for short stature secondary to pituitary-hypothalamic involvement. Bitemporal field defects occur because of pressure or injury to the optic chiasm. The tumor can also compress the third ventricle, causing noncommunicating hydrocephalus. With prominent hydrocephalus, papilledema is evident. Polyuria from diabetes insipidus results from interruption of the supraoptic-hypophyseal tract. Treatment consists of surgical removal of the tumor and hormone replacement. In this patient, Figure A (the sagittal view) shows a large suprasellar mass, and Figure B (the coronal view) shows compression of the pituitary gland and a large suprasellar mass.

(Choice 1) commonly presents with symptoms of increased intracranial pressure (headache, nausea, vomiting, and hydrocephalus) and seizures.

(Choice 2) presents with failure of upward gaze (Parinaud syndrome).

(Choice 4) Other central nervous system (CNS) symptoms are uncommon. Chromophobic adenomas of the pituitary may also cause short stature and diabetes insipidus. Calcifications will not be present.

(Choice 5) is a childhood histiocytosis. Presentation is variable, but skeletal involvement occurs in 80% of patients. Skin lesions, exophthalmos, and hepatosplenomegaly occur in 20% of these patients. Pituitary dysfunction may result in failure to thrive; however, calcifications are not seen on CT.

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2. ☐ A pinealoma
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160. Question

1 points

Category: Pediatrics

A 12-year-old boy is brought to the emergency department with a temperature of 39.1 C (102.4 F) at home, difficulty speaking, and odynophagia for 2 days. Physical examination reveals marked erythema of the right tonsil pillar and edema of the uvula with deviation to the left. In addition to anaerobic bacteria, which of the following organisms is most likely to be isolated from a tonsillar pillar aspirate?

- ☒ Beta-hemolytic Streptococcus
- ☐ Enterococcus
- ☐ Haemophilus influenzae type B
- ☐ Staphylococcus aureus
- ☐ Streptococcus pneumoniae

INCORRECT ❌

The correct answer is 1.

The probable diagnosis is peritonsillar abscess, which is the most common complication of pharyngitis or tonsillitis. Patients usually present with fever, dysphagia, and odynophagia (pain when swallowing). Drooling is common. Physical examination findings include erythema of the involved tonsillar pillar, edema of the pillar and soft palate, deviation of the uvula toward the contralateral side, and trismus. The tonsils may be inflamed and exudative.

(Choices 2,3,4 & 5) The most common organisms isolated from peritonsillar abscesses are β -hemolytic streptococci and anaerobic bacteria. Haemophilus influenzae type b, Moraxella catarrhalis, and Pseudomonas aeruginosa are cultured less frequently. As is typical for most oral infections, mixed cultures are common. Enterococci, Staphylococcus aureus, and Streptococcus pneumoniae generally are not isolated.

Treatment of peritonsillar abscess includes parenteral antibiotics that are active against anaerobes and streptococci, as well as drainage of the abscess. Penicillin remains the drug of choice. In patients who are allergic to penicillin, clindamycin or a cephalosporin can be used as an alternative with the addition of metronidazole. Needle aspiration of the abscess is usually performed in the emergency department. In those in whom needle aspiration fails, subsequent incision and drainage, or tonsillectomy may be necessary.

161. Question**1 points****Category: Pediatrics**

A 5-year-old boy is scheduled for a medical checkup and MMR booster before beginning the school year. All of his vaccinations are up to date, but his mother reports that he had a reaction to the first MMR shot, which was given at 12 months of age. The reaction involved a cough, red eyes, and a rash over the skin that resolved in about 1 week. The mother reports that the child is presently resolving from a "mild case of the flu," with mild fever, coughing, and rhinorrhea, that has kept him awake during the last couple of nights. Past medical history is significant for anaphylaxis to previously administered amoxicillin and neomycin. There is no history of major illnesses. Which of the following is a contraindication to the administration of MMR vaccine in this patient?

1. ☐ Anaphylaxis to amoxicillin
2. ☒ Anaphylaxis to neomycin ✓
3. ☐ Pregnancy in the mother
4. ☐ Previous febrile reaction to the MMR vaccine
5. ☐ Upper respiratory tract infection with low-grade fever

INCORRECT ✗

The correct answer is 2.

It is important to know the contraindications to childhood immunizations. The true contraindications to the MMR vaccine include anaphylactic reaction to the vaccine or any of its components (neomycin, gelatin), immunodeficiency, pregnancy, and untreated and active tuberculosis. Precautions are given in cases of moderate or severe illness, recent administration of immunoglobulin containing blood products, and thrombocytopenia or thrombocytopenic purpura.

(Choice 1) is not a contraindication to MMR vaccination because it is not a component of the vaccine. Components of the vaccination to which patients may be allergic include neomycin and gelatin.

(Choice 3) MMR vaccination is contraindicated in pregnant women due to its possible effects on fetal development. However, MMR vaccination is not contraindicated in persons who are in contact with pregnant women.

(Choice 4) is not a contraindication to vaccination. Some patients can develop measles-like infection with cough, cold, red eyes, and rash. However, it typically lasts 2 to 5 days and is self-limiting. An anaphylactic reaction to a previous vaccination, however, is a contraindication to repeat doses.

(Choice 5) Febrile respiratory illness or any other active febrile infection is not a contraindication. However, the Advisory Committee on Immunization Practices (ACIP) for the Centers for Disease Control (CDC) has recommended that vaccines be administered to persons with minor illnesses, such as diarrhea, mild upper respiratory infection with or without low-grade fever, and other 19–grade febrile illnesses.

162. Question**1 points****Category: Pediatrics**

An 8-year-old female presents to your office because her mother has noticed "several physical changes." She denies headaches, emesis or visual disturbances. Her medical history is significant for two bone fractures which occurred at 6 and 7 years of age. Her family history is unremarkable. She has a healthy 15-year-old brother. She has a moonlike face. Physical examination reveals two light-brown spots with irregular contours on the back of the shoulders and left side of the neck. There is a bruise on the right arm, and elevation of the breasts with budding and enlargement of the areola. There is coarse, dark pubic hair along the labia and the pubic junction. The eye examination is normal. What is the most likely diagnosis?

1. ☐ Von Recklinghausen disease
2. ☒ McCune-Albright syndrome
3. ☐ Peutz-Jeghers syndrome
4. ☐ Sturge-Weber disease
5. ☐ Cushing's disease

INCORRECT ❌**The correct answer is 2.**

McCune-Albright syndrome is a rare condition characterized by precocious puberty, café au lait spots and multiple bone defects (polyostotic fibrous dysplasia). It is responsible for 5% of the cases of female precocious puberty, and may be associated with other endocrine disorders, such as hyperthyroidism, prolactin- or GH-secreting pituitary adenomas, and adrenal hypercortisolism. McCune-Albright syndrome is sporadic and has been recently attributed to a defect in the G-protein *cAMP*-kinase function in the affected tissue, thereby resulting in autonomous activity of that tissue.

(Choice 1) Unlike in Von Recklinghausen syndrome, the café au lait spots of McCune-Albright syndrome are large, have irregular borders and are not associated with axillary or genital freckles.

(Choice 3) Peutz-Jeghers syndrome is characterized by gastrointestinal tract polypoidosis and mucocutaneous pigmentation. It may also involve the development of an estrogen-secreting tumor, leading to precocious puberty.

(Choice 4) Sturge-Weber disease is a sporadic phakomatosis characterized by mental retardation, seizures, visual impairment and a characteristic port-wine stain over the territory of the trigeminal nerve.

(Choice 5) This patient's moonlight facies and bruises indicate Cushing's syndrome, which occurs in association with McCune-Albright syndrome. Cushing's disease consists of hypercortisolism secondary to an ACTH-secreting pituitary tumor.

163. Question

1 points

Category: Pediatrics

A 6-year-old presents to his pediatrician with left ear pain. His parents state that he had been in his usual state of good health until he went swimming in a lake a few days ago. The patient's vital signs are within normal limits for age. He is afebrile and appears nontoxic. Physical examination of the left ear reveals edema and erythema of the ear canal, as well as a greenish otorrhea. It is difficult to visualize the tympanic membrane. Pain is increased by manipulation of the pinna and pressure on the tragus. Which of the following is the most likely pathogen producing this patient's symptoms?

1. ☐ *Haemophilus influenzae*
2. ☒ *Pseudomonas aeruginosa* ✓
3. ☐ *Moraxella catarrhalis*
4. ☐ *Streptococcus agalactiae*
5. ☐ *Escherichia coli*

INCORRECT ✗

The correct answer is 2.

External otitis, or "swimmer's ear," is caused by excessive moisture in the ear canal, which causes the loss of protective cerumen leading to chronic irritation. However, cerumen impaction with trapping of water can also cause infection. The patient presents with ear pain that is worsened by manipulation of the pinna. The ear canal is erythematous and edematous. Otorrhea may be seen. *Pseudomonas aeruginosa* is the most commonly isolated bacterium in external otitis. Other organisms include *Enterobacter aerogenes*, *Proteus mirabilis*, coagulase-negative staphylococci, *Klebsiella pneumoniae*, streptococci, *Staphylococcus aureus*, diphtheroids, and fungi.

(Choices 1 & 3) Both cause otitis media, but not as often as *Streptococcus pneumoniae*.

(Choice 4) has been isolated from middle ear fluid in neonates with otitis media.

(Choice 5) However, both *S. agalactiae* and *Escherichia coli* are leading causes of sepsis and meningitis in neonates.

164. Question

1 points

Category: Pediatrics

A 7-month-old boy is brought to his physician because of increased agitation and restlessness. Lung examination reveals crackles and decreased breath sounds bilaterally. Chest x-ray films are notable for bilateral pneumonia. Arterial blood gas analysis reveals an oxygen tension of 45 mm Hg and a carbon dioxide tension of 60 mm Hg. Which of the following is the most appropriate next step in management?

1. ☐ Obtain blood cultures
2. ☐ Administer oxygen
3. ☐ Administer bronchodilators
4. ☐ Administer antibiotics
5. ☒ Insert endotracheal tube ✓

INCORRECT ✗

The correct answer is 5.

The child is in respiratory failure and needs emergent care. Alveolar ventilation must be restored to lower the carbon dioxide level. The child's restlessness is a clue to the extent of respiratory insufficiency. Intubation will improve ventilation while delivering oxygen.

(Choice 1) will be crucial in guiding antibiotic selection, but these can be obtained after the child is intubated.

(Choice 2) Delivery of oxygen without intubation will not improve the alveolar ventilation to reduce the level of carbon dioxide tension.

(Choice 3) The patient is not wheezing, and bronchodilators will not obviate the need for mechanical ventilation.

(Choice 4) Antibiotics should be administered after the patient is stabilized. Intubation should not be delayed.

165. Question

1 points

Category: Pediatrics

A 7-year-old girl was found in a routine health supervision visit to have bilateral breast tissue development. She also had long, pigmented hair over the labia majora. Her height and weight are both at the 80th percentile for her age. Which of the following is the most appropriate management?

- ☐ CT of the head and abdomen
- ☐ Pelvic ultrasonography
- ☒ Radiography of the head and wrist
- ☐ Reassurance to the parents that it is normal
- ☐ Thyroid stimulating hormone (TSH) level

INCORRECT ❌

The correct answer is 3.

The girl in this clinical vignette has precocious puberty, which is defined as the appearance of pubertal signs before age 8 in girls and before age 9 in boys. In young girls, it involves early development of both breasts and pubic hair. Somatic development also increases to yield a higher-than-average height and weight.

A detailed history should be obtained, including the chronology of secondary sexual development, growth patterns, intercurrent illness, and medication. Physical examination should document the child's Tanner stage of breasts and pubic hair, and growth parameters, and should include neurologic and ophthalmologic examinations. Evaluation of precocious puberty should begin with radiography of the hand and wrist to determine the bone age. If the patient has a normal bone age, she has an incomplete form of sexual precocity. Outpatient follow-up of the patient for 6-12 months is indicated to determine whether complete precocious puberty or another condition is developing. A delayed bone age suggests hypothyroidism. An advanced bone age requires further evaluation.

(Choices 1 & 2) CT scans of the head and abdomen and pelvic ultrasonography are indicated if the bone age is advanced. Ten percent of girls with true precocious puberty have a brain tumor. CT scan of the abdomen and pelvic ultrasonography are helpful in identifying an abdominal mass such as ovarian tumor or adrenal tumor.

(Choice 4) Reassurance to the parents that the early development of both breasts and pubic hair is normal is absolutely inappropriate. Seven years of age is too young for such advanced secondary sexual development.

(Choice 5) should be obtained if the bone age is delayed and precocious puberty secondary to hypothyroidism is suspected. Prolonged hypothyroidism causes growth retardation and delayed bone age. Since TSH and gonadotropins have a similar structure, increased TSH levels in hypothyroidism cause subsequent stimulation of ovaries and testes.

166. Question**1 points****Category: Pediatrics**

A 24-month-old girl is brought to the pediatrician's office for evaluation because her mother noticed a yellowish discharge on the girl's underwear for the past 3 days. She had no fever, but her mother said she has been fussier recently. On physical examination, the girl is appears excessively anxious about contact with the physician. Her introitus is inflamed, and the hymeneal edge is jagged at the 8 o'clock position. A vaginal culture is taken. Which of the following organisms, if isolated from the vaginal vault, would constitute the most definitive evidence of sexual abuse?

1. ☐ *Candida albicans*
2. ☐ *Chlamydia trachomatis*
3. ☐ *Gardnerella vaginalis*
4. ☐ *Pseudomonas aeruginosa*
5. ☒ *Neisseria gonorrhoeae* ✓

INCORRECT ✗**The correct answer is 5.**

The physical examination findings, in particular a posterior tear of the hymen, in this 24-month-old girl are highly suggestive of sexual abuse. However, physical findings alone are not enough to prove a sexual abuse case. Cultures taken from the vaginal vault of the suspected victim are essential in diagnosis and documentation. Of all the pathogens associated with sexual abuse, *Neisseria gonorrhoeae* provides definitive evidence.

(Choice 1) is a common infection of the diaper area for both boys and girls. Vaginal candidiasis can occur in a 24-month-old child.

(Choice 2) can be vertically transmitted to the infant from the mother at birth, and persists for 3 years. It could be associated with sexual abuse but cannot be confirmatory in this case, when it is isolated from a 24-month-old girl.

(Choice 3) presents as vaginitis with discharge. It is uncommon in young girls but does not confirm sexual abuse.

(Choice 4) rarely causes vaginitis and is not associated with sexual abuse.

167. Question

1 points

Category: Pediatrics

A 3-day-old, full-term baby boy is brought into the emergency department because of feeding intolerance and bilious vomiting. X-rays films show multiple dilated loops of small bowel and a "ground glass" appearance in the lower abdomen. The mother has cystic fibrosis. Which of the following diagnostic tests would also have therapeutic value?

1. ☐ Barium enema
2. ☒ Gastrografin enema ✓
3. ☐ Colonoscopy
4. ☐ Endoscopic retrograde cholangiopancreatogram (ERCP)
5. ☐ Full thickness rectal biopsy

INCORRECT ✗

The correct answer is 2.

Anytime you see an infant with abdominal difficulties plus cystic fibrosis, think of meconium ileus. The vignette did not say that this infant had cystic fibrosis (he has not been diagnosed yet; he is only 3 days old), but the mother provides the connection. A Gastrografin enema will show the unused microcolon and the pellets of inspissated meconium higher up. As the hypertonic solution of Gastrografin draws water into the lumen, the inspissated meconium will be diluted and "unplugged." More complete diagnostic steps and treatment for the cystic fibrosis will follow.

(Choice 1) is a tantalizing distracter because it also serves double duty as diagnosis and therapy, but not in this condition. It does so in intussusception, which we see in chubby 2-year-olds with colicky pain and the famous "currant jelly stool," not in the newborn.

(Choice 3) can also do double duty, but it does so in Ogilvie syndrome, the paralytic ileus of the colon that we see in old, immobilized people.

(Choice 4) is also a diagnostic test with therapeutic value, but its role is in clearing up an obstructed biliary tract.

(Choice 5) And as for rectal biopsy, save it for the infant suspected of having aganglionic megacolon; it will give you the diagnosis but will not help with the therapy.

168. Question

1 points

Category: Pediatrics

A 2-month-old girl presents to her pediatrician's office for well-child care. Her mother complains of excessive tearing of the baby's left eye for the past 4 weeks. Each morning, a yellow crusty discharge is noted along the lashes of the left eye. The conjunctiva appears un-inflamed. The right eye is not affected. On physical examination, the infant is otherwise well and achieving adequate weight gain on an exclusive breast milk diet. She is developmentally appropriate, including visually tracking 180 degrees. Which of the following is the most likely diagnosis?

1. ☒ Dacryostenosis ✓
2. ☐ Gonococcal conjunctivitis
3. ☐ Normal infant eye
4. ☐ Viral conjunctivitis
5. ☐ Vitamin A deficiency

INCORRECT ✗

The correct answer is 1.

Dacryostenosis, or congenital nasolacrimal duct obstruction, occurs in at least 6% of all newborns. It is characterized by unilateral tearing and yellow crusting of the eye each morning, sometimes so thick that the infant cannot open the eyelids. Although uncommon, dacryostenosis can be bilateral. Parents are advised to massage the inner canthus of the eye twice daily to encourage resolution of the obstruction. Resolution occurs by 1 year of age in 90% of infants.

(Choice 2) is unlikely because perinatal transmission of gonococcus results in conjunctivitis several days after birth. Clinical findings include lid edema, purulent exudate, and corneal ulcerations. Early diagnosis requires Gram staining of the exudate.

(Choice 3) Eye discharge is never a normal finding in infants, therefore this choice is incorrect.

(Choice 4) is unlikely without symptoms of upper respiratory infection, such as rhinorrhea, cough, fever, lethargy, or poor feeding. Eye involvement may begin unilaterally but progresses to bilateral involvement within 1-2 days. The major clinical finding is an inflamed or red conjunctiva.

(Choice 5) In breast-fed infants, vitamin A deficiency is extremely unlikely. The major clinical finding of avitaminosis is a thick and hazy cornea (keratomalacia). Bottle-fed infants are also unlikely to have vitamin A deficiency, since commercial formulas contain adequate daily allowances of essential vitamins if they are mixed to the correct concentration. Unfortunately, this is not always the case.

169. Question

1 points

Category: Pediatrics

A 2-year-old girl is brought to the physician because of protracted irritability, crying, and loss of appetite. She recently had a sore throat. Her temperature is 38.5 C (101.3 F). Physical examination is unremarkable, except for abnormalities of the tympanic membrane detected on otoscopic examination. Which of the following signs or symptoms correlates best with a diagnosis of acute otitis media?

1. ☐ Color change of tympanic membrane
2. ☐ Fever
3. ☐ Opacification of tympanic membrane
4. ☐ Otalgia
5. ☐ Otorrhea
6. ☒ Reduced tympanic membrane mobility

INCORRECT ❌

The correct answer is 6.

Acute otitis media (AOM) is one of the most frequent conditions affecting infants and young children. Accumulation of exudative fluid within the middle ear results in distention of the tympanic membrane and pain. By the age of 3 years, approximately 50% of children will have had at least one episode. Therefore, all primary care physicians should become skilled in the diagnosis of AOM, which entails a careful otoscopic examination. Physical signs of AOM include bulging and fullness of the tympanic membrane with reduced tympanic membrane mobility appreciated under positive pressure.

(Choice 1) is neither sensitive nor specific of AOM.

(Choice 2) is a nonspecific sign that has little correlation with AOM. Children with AOM often have a concomitant upper respiratory infection that justifies temperature elevation.

(Choice 3) is no longer considered a useful diagnostic criterion for AOM. Opacification of the tympanic membrane, with resultant absent light reflex, may be also observed in normal ears.

(Choice 4) Otalgia, or pain localized to the ear, is absent in a substantial number of cases, especially in infants.

(Choice 5) becomes apparent in only 2% of cases, following spontaneous rupture of the tympanic membrane. One should not await this sign to make a diagnosis of AOM.

170. Question

1 points

Category: Pediatrics

A 15-year-old boy is brought to the pediatrician for a school physical examination. The mother states that they have just moved to this area from Georgia. According to the mother, the child, who is mentally retarded, has been in his usual state of good health. His past medical history is pertinent for a few ear infections and upper respiratory tract infections. Although the mother did not bring the child's previous medical record with her, she believes that the patient's immunizations are current. Pertinent findings on the physical examination include a prominent jaw, protruding ears, Macroorchidism, and developmental delay. Which of the following is the most likely diagnosis?

1. ☐ Klinefelter syndrome
2. ☐ Trisomy 18
3. ☐ Prader-Willi syndrome
4. ☐ Trisomy 21
5. ☒ Fragile X syndrome ✓

INCORRECT ✗

The correct answer is 5.

Associated characteristics of fragile X syndrome include mental deficiency, prominent jaw, large ears with soft cartilage, and Macroorchidism in postpubertal males. Fragile X syndrome is the most common form of inherited mental retardation, affecting approximately 1 in 1,250 males and 1 in 2,000 females. Although inheritance is X-linked, inheritance does not follow classic Mendelian patterns. Most males who carry the mutation are retarded and show the typical phenotypic pattern, whereas about 20% have no obvious symptoms but nonetheless are obligate carriers. Conversely, about 33% of heterozygous females show phenotypic effects, but mental retardation only occurs if the mutation is inherited from the mother. Moreover, the risk of clinical symptoms and the degree of mental retardation increases in succeeding generations. The explanation lies in the fact that a CGG triplet repeat is found in the first untranslated exon; in affected individuals, this repeat is expanded to hundreds or even thousands of repeats. Above a certain critical number, the gene becomes hyper methylated and is shut off. Further variability is built into the condition by the fact that the expanded repeat is unstable during mitosis, causing extensive somatic mosaicism with respect to the number of repeats in various cell types. The diagnosis of fragile X syndrome should be considered in any child with undiagnosed developmental and/or mental retardation. This can now be done using molecular methodologies that are more efficient and easier to do than the classic cytogenetic analyses.

(Choice 1) In patients with Klinefelter syndrome, hypogonadism, long extremities, decreased intelligence, and behavioral problems can be seen.

(Choice 2) Clinical features of trisomy 18 are a prominent occiput and low-set, posteriorly rotated, malformed auricles; clenched hand with overlapping fingers; and rocker-bottom feet.

(Choice 3) The physical features associated with Prader-Willi syndrome include a narrow face, almond-shaped eyes, a thin upper lip, hypoplastic gonads, small hands and small feet, and truncal obesity after 1–4 years of age. These patients also have slow motor development, delayed puberty, mental retardation, and behavioral problems.

(Choice 4) Physical manifestations of trisomy 21 include epicanthal folds, Brushfield's spots, and simian crease, a wide space between the first and second toes, a short fifth finger, and small ears.

171. Question**1 points****Category: Pediatrics**

A previously healthy 3-year-old boy brought in by his parents for help with potty training. The boy refuses to use the toilet. If his parents try to place him on the toilet, he becomes upset and cries. He has regular urination in his diaper. He has 1-2 soft bowel movements a day. In the office, you note that the child speaks in short sentences that are mostly understandable. He runs well and can climb up on the examination table without help. His physical examination is unremarkable. Which of the following is the next best step in the management of this patient?

1. ☐ Increase fiber in his diet
2. ☐ Put him on the toilet at regularly scheduled times
3. ☐ Use a positive reward system
4. ☒ Stop potty training attempts for several months ✓
5. ☐ Perform a urinalysis

INCORRECT ✗**The correct answer is 4.**

Toilet training usually occurs between the ages of 2-4 years old. Parents should be counseled that several milestones must be met by the child to become potty trained. Children must be aware of bladder filling, be able to consciously tighten the external sphincter, have normal bladder growth, and be motivated to stay dry. They must also have met motor milestones to be able to walk to the toilet, sit upright on the toilet, and the language to indicate the need to use the bathroom. The child will demonstrate readiness to begin toilet training when they communicate the need to urinate or defecate prior to passing urine or stool, and when they can withhold the urine or stool for a brief period of time.

When a child is ready, toilet training consists of placing the child on the toilet at regular times and using positive reinforcement as a reward. Girls are usually toilet trained faster than boys, but 90-95% of child will achieve daytime continence by 5 years old. When a child is not ready for toilet training, such as the child in the vignette, forcing him to sit on the toilet will make him more reluctant and can make potty training very difficult. The best step in the management of a reluctant child is to stop toilet training for several months and allow the child to become interested on his own.

(Choice 1) The child is having 1-2 soft stools a day so constipation is not a concern. Therefore, increasing fiber will not help the child become toilet trained.

(Choices 2 & 3) Placing the child on the toilet at regular times and using positive reward systems are the correct steps to use to potty train a child. However, the child must show a willingness to be potty trained before these methods will work.

(Choice 5) The child has no symptoms of a urinary infection so a urinalysis is not indicated.

172. Question

1 points

Category: Pediatrics

A 3-year-old boy is brought to the emergency department because of a worsening cough over the past week. His temperature is 38.9 C (102 F), and inspiratory stridor is noted. A plain film of the neck reveals subglottic swelling. He is noted to have copious thick secretions and a barking cough. He has not had such events previously, and his parents deny recent contact with sick children. The patient is in respiratory distress and is noted to be retracting his subcostal muscles to breathe. Which of the following is the next most appropriate step in management?

1. ☐ Administer albuterol
2. ☒ Administer racemic epinephrine ✓
3. ☐ Administer corticosteroids
4. ☐ Administer N penicillin
5. ☐ Endotracheal intubation

INCORRECT ✗

The correct answer is 2.

The patient has infectious laryngotracheitis (viral croup), which is caused by the parainfluenza virus. Symptoms are often worse at night and include a characteristic barking cough. Fever is usually low grade, but temperatures as high as 104 F has been noted. Epiglottitis is in the differential but is rapidly progressive and is marked by the abrupt onset of high fever. Racemic epinephrine by aerosol has been shown to provide symptomatic relief by vasoconstriction and reduction of local edema. This drug should be used in the moderately ill patient since it may eliminate the need for intubation.

(Choice 1) would help in treating the bronchospasm but will be of little use in treating croup. In mild illness, humidification may help.

(Choice 3) Some data support the use of corticosteroids in severely ill patients, although this remains controversial. Patients given dexamethasone with a repeat dose in 2 hours have had a shorter course of illness than those not given steroids.

(Choice 4) This is a viral illness, and antibiotics will be of little utility. However, acute epiglottitis is caused by *Haemophilus influenzae* type B, and more recently, Group A β -hemolytic streptococci. A third-generation cephalosporin would be appropriate therapy until culture results are known.

(Choice 5) may be needed if the respiratory distress worsens and epinephrine and steroids fail.

173. Question

1 points

Category: Pediatrics

A 10-year-old girl is brought to a pediatrician because her mother notices that she stumbles frequently at night, even with adequate lighting. Visual field testing demonstrates a relatively narrow mid-peripheral ring scotoma. Ophthalmoscopy demonstrates dark pigmentation in a bone spicule configuration involving the equatorial retina. Additional findings include a waxy yellow appearance to the disk and narrowed retinal arteries. Which of the following is the most likely diagnosis?

1. ☐ Cataract
2. ☐ Central retinal artery occlusion
3. ☐ Retinal detachment
4. ☒ Retinitis pigmentosa ✓
5. ☐ Uveitis

INCORRECT ✗

The correct answer is 4.

Retinitis pigmentosa is an often hereditary progressive degenerative disease of the retina. Patients present with progressive night blindness, visual field constriction with a ring scotoma, and loss of acuity. Characteristically, the rods in the equatorial retina are affected first, producing a mid-peripheral ring scotoma that can be detected with visual field testing. The ophthalmoscopic findings illustrated in the question stem are typical; additional manifestations include cataracts, myopia, and opacities in the vitreous humor. Some patients also have hearing loss. The condition is slowly progressive (usually leading to complete blindness over a period of several decades) and is at the moment untreatable, although fetal retinal transplants show some promise.

(Choices 1 & 2) A cataract is an opacity of the lens of the eye. Central retinal artery occlusion causes sudden, painless, unilateral blindness, and is associated with pallor of the macula.

(Choice 3) In the case of retinal detachment, ophthalmoscopy reveals a raised area in one retina and would not selectively affect night vision.

(Choice 5) is an inflammation of the iris, ciliary body, or choroid, and may cause visual loss secondary to "floaters" in the vitreous humor.

174. Question

1 points

Category: Pediatrics

Parents present to the emergency department carrying their 4-year-old child, who is lethargic and has excessive oral secretions, miosis, tearing, and "soiled" trousers from urination and defecation. Remnants of emesis are seen on his clothing. Tremors, fasciculations, and hypertension are also present. The parents tell the physician that the child had been in his usual state of good health until that afternoon. The patient's symptoms developed while playing in a field recently sprayed with insecticides. The physician suspects organophosphate poisoning. To treat the nicotinic effects, the physician should use which of the following?

1. ☐ Atropine
2. ☐ British antilewisite
3. ☒ Pralidoxime ✓
4. ☐ Calcium disodium ethylenediaminetetraacetate
5. ☐ Naloxone

INCORRECT ✗

The correct answer is 3.

The specific antidote for nicotinic manifestations of organophosphate toxicity is pralidoxime. It restores the activity of acetylcholinesterase. Nicotinic effects for organophosphates include fasciculations, twitching, weakness, areflexia, tachycardia, and hypertension.

(Choice 1) is an antidote for the muscarinic effects of organophosphate toxicity. Some muscarinic effects include salivation, lacrimation, urination, defecation, and abdominal cramps.

(Choices 2 & 4) British antilewisite (BAL) and calcium disodium ethylenediaminetetraacetate (Ca-EDTA) are used for the treatment of lead toxicity. These therapies are used in patients with a venous blood lead level concentration of 70 µg/dL or above. Ca-EDTA and BAL are used in combination for children with encephalopathy or evidence of encephalopathy.

(Choice 5) is used as an antidote for opioid poisoning.

175. Question**1 points****Category: Pediatrics**

An 8-year-old boy is brought to his pediatrician by his mother, who disclosed that the patient had been in his usual state of good health until this morning, when he awoke covered with bruises and purple dots. The patient has no history of trauma, and there is no history of bleeding disorders in the patient or family. The past medical history is only significant for a mild upper respiratory infection 1 week ago. The pediatrician notes that the boy has petechiae and purpura over his entire body. A complete blood count (CBC) is obtained. The platelet count is 20,000 platelets/ μL (normal is 150-450 $\times 10^3/\mu\text{L}$), but the rest of the CBC results are normal. No hepatosplenomegaly is present. A bone marrow aspirate exhibits normal megakaryocytes, but platelet budding is poor. Which of the following is the most appropriate approach to the management of this patient?

1. ☐ Platelet concentrates should be given prophylactically to prevent bleeding.
2. ☐ Whole blood should be administered to prevent bleeding.
3. ☒ γ -Globulin and/or corticosteroid should be administered. ✓
4. ☐ Vitamin C should be administered.
5. ☐ Vitamin K should be administered.

INCORRECT ✗

The correct answer is 3.

This patient has idiopathic (autoimmune) thrombocytopenic purpura (ITP). ITP is the most common thrombocytopenic purpura of childhood. The disease usually follows a viral infection. The infection seems to trigger an immune mechanism that starts platelet destruction. This destruction is manifested as sudden onset of bruising and petechiae. Other than this condition, the patient looks well, but bleeding into tissues can occur. Platelet counts are depressed and bleeding time is prolonged, but the white blood cell (WBC) count is normal. Platelet antibodies are commonly seen. Bone marrow examination reveals normal megakaryocytes. The disease is self-limited, with resolution of the petechiae over 1 or 2 weeks, although thrombocytopenia may persist longer. The prognosis for ITP is excellent. Most patients recover within 2 or 3 months. Approximately 90% of children regain normal platelet counts within 9-12 months. In cases of mild thrombocytopenic purpura, with no hemorrhages of the retina or mucous membranes, therapy may not be appropriate. However, therapy with intravenous (IV) γ -globulin infusions and/or corticosteroid therapy can be beneficial. IV γ -globulin infusions induce a sustained rise in platelet counts, while corticosteroid therapy reduces the severity and shortens the duration of the initial phase.

(Choice 1) Platelet concentrates are recommended only in life-threatening situations because their short lifespan is reflected by the temporary nature of the rise in platelet count.

(Choice 2) Therefore, it is not necessary to administer whole blood, which should only be used emergently for hypovolemia due to blood loss.

(Choices 4 & 5) Vitamins C and K will not ameliorate the disease. The patient's complete blood count (CBC) is normal except for the platelet count.

176. Question

1 points

Category: Pediatrics

A 3-year-old girl with a ventricular septal defect (VSD) presents to the emergency department after a 15-minute focal seizure of her left arm and leg. A brief history reveals that the child has no known seizure disorder and has been having a low-grade fever at home for about 4 days. She also has been less active and has had poor appetite. On physical examination, her temperature is 40.2 C (104.3 F), and her pulse is 82/min. She is not responsive to her name, but she is responsive to painful stimuli with withdrawal of her extremities. Cardiac examination is significant for a grade 3 systolic murmur best heard at the left lower sternal border. Neurologic examination reveals anisocoria with a dilated right pupil. After stabilization, which of the following is the most appropriate next step in diagnosis?

1. ☒ CT of the brain ✓
2. ☐ ECG
3. ☐ Electroencephalography
4. ☐ MRI of the brain
5. ☐ Complete blood count and blood culture
6. ☐ Lumbar puncture

INCORRECT ✗

The correct answer is 1.

A child with a known cardiac defect now presenting with fever, seizure, and focal neurologic deficit (anisocoria) is likely to have a brain abscess. Ventricular septal defect (VSD) predisposes the patient to an occult infection in the heart, i.e., endocarditis. Vegetations usually develop around the defect. Seeding of the bacteria may occur, and the patient thus becomes bacteremic. The bacteria may travel to the brain and cause a brain abscess. Common pathogens include *Streptococcus viridans*, *Bacteroides*, *Enterobacteriaceae* and *Staphylococcus aureus*. Brain abscess is a life-threatening condition that requires rapid recognition and treatment. CT of the brain is the best diagnostic test for brain abscess. Surgical drainage and IV antibiotics are the treatment of choice. Broad spectrum antibiotics, such as a third generation cephalosporin with metronidazole, are necessary. Oxacillin or vancomycin can be used if *S. aureus* is present. If CT scan reveals that the abscess is less than 2.5 cm and the patient is neurologically stable and conscious, it is recommended to just start antibiotics and observe. Otherwise, surgical drainage is necessary.

(Choice 2) is not helpful in the diagnosis of this child. Even though she has a cardiac condition (VSD), it is apparent that her clinical deterioration is secondary to a serious infection and a neurologic condition, rather than an abnormal heart rhythm.

(Choice 3) is an appropriate initial diagnostic procedure for this critically ill child. It may help to elucidate the kind of seizure that she had. It is not immediately necessary, however, because the confirmation of a brain abscess by a CT scan is more crucial for prompt diagnosis and intervention.

(Choice 4) can show fine details of the soft tissue. In this case of brain abscess, however, a rapid study such as the CT scan is more appropriate because it is faster and easier to obtain and is adequate to diagnose a brain abscess.

(Choice 5) is helpful but is not diagnostic of brain abscess. An elevated white count is not always present. Multiple blood cultures should be obtained but they are not as helpful as the CT scan.

(Choice 6) is contraindicated in this case because the child might have increased intracranial pressure. Brain stem or uncal herniation might occur.

178. Question

1 points

Category: Pediatrics

A 2-year-old girl is brought to the emergency department with a fever, chills, poor appetite, and vomiting. On examination, she is irritable and diaphoretic. Her temperature is 39.2 C (102.5 F), blood pressure is 80/48 mm Hg, pulse is 88/min, and respirations are 17/min. She is tender at the left costovertebral angle. Initial laboratory tests show the following:

Leukocyte count: 16,300/mm³

Hemoglobin: 12.5 g/dL

Platelet count: 245,000/mm³

Blood urea nitrogen: 6mg/dL

Creatinine: 0.5 mg/dL

Urinalysis is positive for leukocyte esterase and nitrite, with 150 white blood cells/hpf. After IV antibiotic administration and stabilization, what is the most appropriate diagnostic study?

1. ☐ CT of the abdomen and pelvis
2. ☐ IV pyelography
3. ☐ Plain abdominal radiography
4. ☐ Radionuclide imaging of the kidneys
5. ☒ Voiding cystourethrography ✓

INCORRECT ✗

The correct answer is 5.

This girl has pyelonephritis, which is an upper urinary tract infection (UTI) that involves one or both of the kidneys. Children with pyelonephritis present with high fever, dysuria, vomiting, abdominal pain, back pain, and irritability. They often have costovertebral tenderness on examination. The laboratory evaluation often shows leukocytosis. Urinalysis shows the presence of leukocyte esterase, nitrite, and increased white blood cells (>5/hpf). It is also important to note the number of squamous epithelial cells. The urine specimen is considered contaminated and is a poor specimen if there are more than 5 squamous epithelial cells per hpf. Obtaining a urine culture is also important to identify the pathogen and its susceptibility to different antibiotics. In very young children, a clean catch may not be the best method to obtain a good urine sample; straight bladder catheterization may be necessary. The most common organisms are *Escherichia coli*, *Proteus*, *Klebsiella*, and *Enterobacter* sp. Voiding cystourethrography (VCUG) is indicated in all girls younger than 5 years with the first episode of UTI, because 25 to 30% of such patients have vesicoureteral reflux. Older girls should be evaluated by VCUG after the second episode of UTI. VCUG is also indicated in all boys who have an episode of UTI regardless of age.

(Choice 1) is indicated when there is a suspicion of structural abnormalities of the kidneys, but not for routine evaluation of UTI.

(Choice 2) is used to visualize the renal calyces, ureteral anatomy, urinary diversions, and ectopic ureterocele. It is not routinely recommended after UTI.

(Choice 3) has no role in the evaluation of a known UTI.

(Choice 4) involves the administration of a radioactive material, usually ⁹⁹Tc-DTPA, to assess the blood flow and excretory function of the kidney. Areas of differential function or obstruction can be detected.

179. Question

1 points

Category: Pediatrics

A nurse notices that a 1-week-old, premature infant in the neonatal unit is experiencing migratory jerks of the extremities. She picks the infant up and can feel that the muscle jerks continue to happen, even when she holds an involved extremity still. After about 5 minutes, the jerking movements stop. Which of the following is the most appropriate first step in diagnosis?

1. ☐ CT scan of head
2. ☐ Electroencephalography
3. ☒ Serum chemistries ✓
4. ☐ Skull x-rays
5. ☐ Ultrasound of head

INCORRECT ✗

The correct answer is 3.

The infant is having neonatal seizures, which can take a variety of forms. In addition to the migratory clonic jerks described in the question stem, these infants may alternatively experience alternating hemi seizures or primitive subcortical seizures. The later may take the form of respiratory arrest, chewing motions, eye deviations, or changes in muscle tone. True grand mal seizures are usually not seen, possibly because the immaturity of the brain makes it difficult to transmit a seizure to all sites. Neonatal seizures can be seen in a variety of settings, which can be roughly sub-classified as primary CNS versus metabolic. The metabolic causes are common, and screening chemistry studies can identify potentially correctable hypoglycemia, hypocalcemia, hypomagnesemia, hypematremia, or hyponatremia. Consequently, the earliest diagnostic step is to order these appropriate serum screening tests, usually on a STAT basis.

(Choices 1,2,4 & 5) CT scan of head, EEG, skull x-rays, and ultrasound of the head can all be very useful in offering diagnostic clues to true CNS problems, but should be deferred until after the child's metabolic status is established.

180. Question

1 points

Category: Pediatrics

Soon after birth, a term newborn is noted to be cyanotic with feeding. The infant was born to a 24-year-old primigravida who had excellent prenatal care. There is no history of tobacco, alcohol, or illicit drug use. Review of the mother's medical record shows that results of all diagnostic studies for sexually transmitted diseases have been negative. The delivery was uncomplicated. The infant weighed 6 lb 13 oz (3.1 kg), and Apgar scores at 1 and 5 minutes were 8 and 9, respectively. Examination of the newborn reveals that crying relieves the cyanosis. Which of the following is the most likely diagnosis?

1. ☐ A tracheoesophageal fistula
2. ☐ Bronchopulmonary dysplasia
3. ☐ Respiratory distress syndrome
4. ☒ Choanal atresia
5. ☐ A patent ductus arteriosus

INCORRECT ❌

The correct answer is 4.

Choanal atresia should be suspected in any infant who develops cyanosis that occurs with feedings or at rest but is relieved by crying. The cyanosis occurs because newborns are obligate nose breathers, and this congenital anomaly consists of a unilateral or bilateral bony or membranous septum between the nose and the pharynx. Diagnosis is made by failure to pass a catheter through the nasal passages.

(Choice 1) is characterized by choking with feeding. An oro-gastric tube curls up and does not pass to the stomach.

(Choices 2 & 3) (Choice 2) is a complication of respiratory distress syndrome (Choice 3) and appears later in the infant's life. Cyanosis is not relieved by crying in either entity.

(Choice 5) is a left-to-right shunt and is not associated with cyanosis.

181. Question

1 points

Category: Pediatrics

A premature infant has a difficult delivery with episodes of arrhythmia and suspected hypoxia ischemia. After the delivery, the infant is lethargic and has periods of apnea. Intracranial hemorrhage is suspected. No obvious head trauma is noted. Cranial ultrasound identifies blood within the ventricles. Which of the following structures is the most likely source of the hemorrhage?

1. ☐ Bridging veins of the skull
2. ☐ Cerebral cortex
3. ☒ Germinal matrix 
4. ☐ Middle meningeal artery
5. ☐ Thalamus

INCORRECT 

The correct answer is 3.

Intraventricular and/or intraparenchymal intracranial bleeding is a common serious problem among newborns, particularly those born prematurely. This diagnosis should be suspected in infants with apnea, seizures, lethargy, or an abnormal neurologic examination. CT scan or ultrasound of the head can confirm the diagnosis. The usual source of the bleeding is the germinal matrix (a mass of embryonic tissue near the caudate nucleus that is not present in adults), which often has been damaged by episodes of hypotension or hypoxia/ischemia. Depending on the exact sites that start to bleed, the blood may either flow into the ventricles or spread into the adjacent brain tissue, "dissecting" a path (and rupturing neuron axons as it goes). This latter form of hemorrhage, which is considered the most serious (and potentially fatal) form of intracranial hemorrhage in infants, typically develops in the first 3 days of life. Infants with small intraventricular hemorrhages often do well; those with large hemorrhages may either die or be left with severe neurologic defects.

(Choice 1) can be a source of bleeding and cause subdural hemorrhage. These vessels are damaged in shaken baby syndrome.

(Choice 2) may be damaged by dissecting blood but is not the usual site of initial bleeding in these infants.

(Choice 4) Bleeding from the middle meningeal artery results in an epidural hematoma. This typically follows trauma. It is, however, an unusual source of bleeding in newborns.

(Choice 5) The thalamus and other areas in the deeper aspects of the brain can be damaged by dissecting blood but are not the usual site of initial bleeding in these infants.

182. Question

1 points

Category: Pediatrics

A 3-year-old girl is believed to have swallowed a marble. She presents to the emergency department unable to speak and begins to become cyanotic. Initial attempts at endotracheal intubation are unsuccessful. Which of the following is the most appropriate next step in management?

- ☐ Continued attempts at endotracheal intubation
- ☐ Cricothyroidotomy (surgical)
- ☐ Face mask 100% O₂ with succinylcholine
- ☐ Formal tracheostomy
- ☒ Needle cricothyroidotomy

INCORRECT ❌

The correct answer is 5.

In any emergency situation, the first step is to secure an airway and verify breathing (the ABCs: airway, breathing, circulation). Because endotracheal intubation was unsuccessful, a surgical airway must be secured. There are two types of cricothyroidotomy: needle and surgical. Needle cricothyroidotomy is the procedure of choice in children younger than 12 years. A patient can be ventilated for 30 minutes this way, allowing time for a more secure airway. In addition, enough pressure may be generated to expel a foreign body in the glottic area.

(Choice 1) Intubation has already failed, and the patient is becoming cyanotic and needs oxygenation.

(Choice 2) Surgical cricothyroidotomy is reserved for patients older than 12 years.

(Choice 3) A face mask with 100% O₂ plus succinylcholine is incorrect because there is complete obstruction, which must be bypassed for oxygenation to be successful.

(Choice 4) is reserved for longterm management of the airway. In this acute setting, a rapid, temporary airway is essential.

183. Question

1 points

Category: Pediatrics

An 18-month-old boy is brought to the office for a well baby visit. The mother appears concerned because a day after the administration of the previous dose DTaP vaccine, the baby developed a seizure which lasted for about 2 minutes. She took the baby to the emergency department, where the baby was kept under observation and subsequently discharged. The baby has had no recurrences, and has been in good health ever since. The child is due for the fourth dose of DTaP vaccine today. Which of the following is the most appropriate course of action?

1. ☐ Do not administer the vaccine
2. ☐ Administer the routine DTaP
3. ☒ Administer the diphtheria and tetanus toxoids, avoid the pertussis component 
4. ☐ Administer the pertussis and tetanus toxoids, avoid the diphtheria component
5. ☐ Administer the diphtheria and pertussis toxoids, avoid the tetanus component

INCORRECT

The correct answer is 3.

The adverse reactions to the DTaP vaccine are usually attributed to the pertussis component of the vaccine. Mild reactions consist of local redness and swelling, irritability, and fever 3 hours and a temperature of $> 105^{\circ}\text{F}$ (40.6°C). An immediate anaphylactic reaction, an encephalopathy, or any CNS complication within 7 days of administration of the vaccine is a contraindication for further administration of DTaP. In these instances, OT should be substituted for DTaP.

(Choices 1, 2, 4 & 5) Since most adverse reactions to the vaccine are due only to the pertussis component (and not due to tetanus and diphtheria toxoids), it is inappropriate to withhold the tetanus and diphtheria vaccine.

184. Question

1 points

Category: Pediatrics

A 1-month-old, previously healthy infant develops forceful projectile vomiting. No bile is seen in the vomitus. After the infant feeds, gastric peristaltic waves are visible crossing the epigastrium from left to right. Several minutes later, the projectile vomiting occurs. Which of the following is the most likely diagnosis?

1. ☐ Diaphragmatic hernia
2. ☐ Duodenal atresia
3. ☐ Esophageal atresia
4. ☒ Hypertrophic pyloric stenosis ✓
5. ☐ Meconium plug syndrome

INCORRECT ✗

The correct answer is 4.

This infant has hypertrophic pyloric stenosis, which is characterized by obstruction of the pyloric lumen by pyloric muscular hypertrophy.

The hypertrophy develops after birth, and typically presents at 4-6 weeks of life. The presentation illustrated in the question stem is typical.

The diagnosis can be confirmed by palpation of a 2- to 3-cm C "walnut-sized" movable mass deep in the right side of the epigastrium.

Ultrasound can also demonstrate the hypertrophied pyloric muscle. Surgical correction is with pyloromyotomy, in which the gastric mucosa is left intact but the muscles of the pyloric areas are incised.

(Choice 1), in which loops of bowel protrude into the chest cavity, produces respiratory distress with bowel sounds heard over the chest.

(Choice 2) also causes projectile vomiting in infants, but the vomitus is bile stained.

(Choice 3) causes regurgitation on feeding but not projectile vomiting.

(Choice 5) causes bowel obstruction in neonates.

185. Question

1 points

Category: Pediatrics

A 5-year-old child develops left-sided ear pain, but her mother is too busy to take her to the pediatrician. There is no improvement in the child's condition, and the mother has noticed painful, swollen, red areas behind the pinna. At this point, the child is brought to the emergency department, where the physician notes the presence of a creamy discharge in the left ear canal and a temperature of 38.6 C (101.5 F). Tuning fork tests localize hearing better on the left ear when placed on apex of head, and bone conduction is greater than air conduction in the left ear. Which of the following is the most likely diagnosis?

1. ☒ Acute mastoiditis ✓
2. ☐ Barotitis media
3. ☐ Cholesteatoma
4. ☐ Chronic otitis media
5. ☐ Meniere disease

INCORRECT ✗

The correct answer is 1.

This child has acute mastoiditis, which can complicate untreated (and occasionally treated) cases of acute otitis media, especially when caused by bacteria. This is a dangerous complication because the mastoid process is adjacent to the brain, and this destructive infection can spread to cause a purulent meningitis. CT scans can be very helpful in defining the extent of the infection. Initial treatment is with intravenous forms of the antibiotics used to cover acute otitis media; coverage is then switched after cultures demonstrate specific antibiotic sensitivities.

(Choice 2) is middle ear damage caused by rapid pressure changes, as in airplane descent or deep sea diving.

(Choice 3) is a benign condition in which stratified squamous keratinizing epithelium grows in the middle ear. It results in a smelly discharge from the ear, hearing loss, vertigo, headache, and facial nerve palsy. Local expansion may result in damage to adjacent structures, and the condition can be fatal if left untreated.

(Choice 4) is the term used for chronic ear drum perforation and the infection that accompanies it.

(Choice 5) causes the cluster of vertigo, tinnitus, and fluctuating hearing loss.

186. Question

1 points

Category: Pediatrics

An 8-year-old boy is brought to the emergency department with a head injury. He hit his head on the ground when he fell off his bicycle. He was not wearing a helmet at the time. There was no loss of consciousness. He vomited 2 times after the accident and now complains of a right-sided headache and inability to hear with his right ear. There is no photophobia or diplopia. On physical examination, his blood pressure is 110/72 mm Hg, pulse is 104/min, and respirations are 22/min. He is alert and oriented and responds appropriately to questions. There is a round hematoma on the right side of his head.

Bloody drainage is noted from his right ear. Which of the following injury is most consistent with these findings?

1. ☐ Concussion
2. ☐ Epidural hematoma
3. ☐ Subdural hematoma
4. ☒ Temporal bone fracture ✓
5. ☐ Tympanic membrane perforation

INCORRECT ✗

The correct answer is 4.

Bleeding from the ear following an injury to the skull is pathognomonic of a temporal bone fracture. The bleeding can be medial to an intact tympanic membrane, from the middle ear through a rupture of the tympanic membrane, or from a fracture line in the ear canal. Hemotympanum gives the tympanic membrane a blueback color. Usually, there is a communication with the subarachnoid space through the fracture line. Often, there is cerebrospinal fluid otorrhea. Cleaning of the ear canal should be avoided for fear of introducing microorganisms. The immediate danger to the patient is the development of meningitis. Prophylactic antibiotic therapy has to be initiated immediately.

Most fractures of the temporal bone are longitudinal (80%) to the long axis of the petrous pyramid. Only 20% are transverse. Longitudinal fractures usually extend through the middle ear into the ear canal, causing rupture of the tympanic membrane. Approximately 35% of longitudinal fractures produce a sensorineural hearing loss, and approximately 15% produce facial paralysis. Transverse fractures extend across the cochlea and fallopian canal, causing a profound, permanent sensorineural hearing loss and a facial paralysis. These fractures are usually well demonstrated with CT.

(Choice 1) is a syndrome of alteration of consciousness secondary to head injury, typically brought about by a sudden change in the momentum of the head. Its usual manifestations include loss of consciousness, temporary respiratory arrest, and loss of reflexes.

(Choice 2) Epidural hematomas are blood clots that form between the inner table of the skull and the dura. Eighty percent are associated with skull fractures across the middle meningeal artery or across a dural sinus. The high arterial pressure of the bleeding vessel dissects the dura away from the skull, permitting the formation of the hematoma. The incidence of skull fractures in children with epidural hematomas is lower than in adults because of the elasticity of the skull.

(Choice 3) usually results from trauma involving acceleration or deceleration head injury and is commonly associated with parenchymal brain injury.

(Choice 5) most commonly happens after an object is inserted into the ear canal and perforates the eardrum.

187. Question

1 points

Category: Pediatrics

A 17-year-old boy is brought to the emergency department by his father after the boy began threatening him at home. Over the last several months, the father reports, the boy has been increasingly abusive. He was recently involved in a fist fight at school. The boy states that there is nothing wrong. He is otherwise healthy. He denies alcohol use, but does admit to occasional marijuana use. On examination he has acne on his forehead and back and his hairline is receding. There is palpable tissue underneath his nipples bilaterally. Heart and lung exams are normal. What substance is this boy most likely abusing?

1. ☒ Anabolic steroids ✓
2. ☐ Cocaine
3. ☐ Heroin
4. ☐ Methamphetamine
5. ☐ Phencyclidine

INCORRECT ✗

The correct answer is 1.

This patient has increased aggression, acne, accelerated male pattern baldness, and gynecomastia concerning for anabolic steroid use. Anabolic steroids are popular among young people and athletes looking to improve their physical appearance and performance. Unfortunately, anabolic steroids (e.g. testosterone, stanozolol, nandrolone) are associated with a significant number of side effects. They impair endogenous testicular function and decrease testicular size and sperm count. Hepatic dysfunction may occur and patients can develop erythrocytosis due to androgen-stimulate erythropoiesis. HDL levels decrease, and high doses may even cause cardiac dysfunction. Psychiatric disorders, like increased aggression and major mood disorders are more common in patients taking anabolic steroids. Gynecomastia can develop because testosterone is converted to estrogen and will drive breast enlargement. Skin changes include acne and premature or accelerated male pattern baldness. Women may have other signs of virilization like hair growth and clitoral enlargement.

(Choice 2) Acute cocaine intoxication is associated with euphoria but can be complicated by aggression, psychiatric changes, and multiorgan dysfunction. Chronic use is associated with cognitive impairment and mood disorders. It is not associated with acne or gynecomastia.

(Choice 3) Heroin intoxication causes symptoms ranging from mild euphoria to stupor and coma. It is associated with respiratory depression, pupillary constriction, and decreased bowel function. Withdrawal might be associated with agitation but not acne or gynecomastia.

(Choice 4) Methamphetamine is a CNS stimulant associated with a number of complications, including hypertension, tachycardia, severe agitation or aggressiveness, and psychosis. Patients might have acne or facial sores due to picking. Male pattern baldness may be seen; however, gynecomastia is absent.

(Choice 5) Phenylcyclidine (PCP) causes brief dissociative psychotic episodes. Patients taking PCP are notoriously aggressive and unaware of pain, but they do not develop acne, baldness, or gynecomastia.

188. Question**1 points****Category: Pediatrics**

A 3-year-old child with a long history of sinopulmonary disease is noted to have difficulty walking and dilated vessels in the conjunctiva and skin. In addition, the skin has lost its normal elasticity. Chest radiology reveals an absent thymic shadow. Pertinent laboratory data reveal elevated u-fetoprotein (AFP) levels, decreased serum immunoglobulin G (IgG) levels, and low IgA and IgE levels. Which of the following is the most likely diagnosis?

1. ☐ Wiskott-Aldrich syndrome
2. ☒ Ataxia-telangiectasia ✓
3. ☐ Rendu-Osler-Weber disease
4. ☐ Friedrich's ataxia
5. ☐ Cerebral palsy

INCORRECT ✗**The correct answer is 2.**

Ataxia-telangiectasia is an autosomal recessive disorder and the most common of the degenerative ataxias. The ataxia usually begins at approximately 2 years of age. It is progressive until full loss of ambulation occurs by adolescence. Telangiectasia (red spots on the skin due to dilated superficial blood vessels) is seen in the eyes, bridge of the nose, ears, and exposed surfaces of the extremities. The skin also loses elasticity. Sinopulmonary infections occur frequently, secondary to abnormal immune function. Low immunoglobulin A (IgA) and IgE levels are seen. The u-fetoprotein (AFP) level is elevated. Children with ataxia-telangiectasia have a much higher risk of developing lymphoreticular and brain tumors.

(Choice 1) is an X-linked recessive syndrome that presents with atopic dermatitis, thrombocytopenic purpura, and an increased susceptibility to infection.

(Choice 3) Rendu-Osler-Weber disease (also known as hereditary hemorrhagic telangiectasia) is characterized by angiomas of the skin, mucous membranes, gastrointestinal tract, and liver, as well as by pulmonary arteriovenous fistulas.

(Choice 4) presents before 10 years of age with progressive ataxia, dysarthric speech, nystagmus, and skeletal abnormalities.

(Choice 5) is a static encephalopathy that is a non progressive disorder of posture and movement. Cerebral palsy may be associated with mental retardation, epilepsy, and behavioral abnormalities.

189. Question

1 points

Category: Pediatrics

A 24-hour-old male Caucasian infant has developed bilious vomiting over the last 3 hours. He hasn't passed his stools yet. His mother is a smoker, and her pregnancy was complicated by polyhydramnios. He was born at term via vaginal delivery, and his APGAR scores were 7 and 9, at 1 and 5 minutes, respectively. His mother started to breastfeed him soon after birth. An uncle on his father's side has cystic fibrosis. His pulse rate is 120/min. Examination reveals a crying infant with a distended abdomen. X-rays of the abdomen reveal a dilated small bowel and a granular, ground glass appearance in the lower abdomen. What is the most likely diagnosis of this patient?

1. ☐ Intussusception
2. ☐ Meconium ileus
3. ☐ Meconium plug syndrome
4. ☒ Duodenal atresia
5. ☐ Esophageal atresia

INCORRECT ❌

The correct answer is 4.

The patient has a very typical presentation of meconium ileus, which is characterized by bilious vomiting, failure to pass meconium at birth, history of polyhydramnios, family history of cystic fibrosis, and a typical appearance on abdominal x-rays. This condition is diagnostic of cystic fibrosis. Although this patient hasn't been screened for cystic fibrosis yet, the diagnosis is very likely, especially with his father's family history. One of the possible complications of meconium ileus is intestinal perforation. If the perforation occurs after birth, pneumoperitoneum will be seen on abdominal x-rays. On the other hand, if it occurs before birth, intraabdominal calcifications will be seen.

(Choice 3) Meconium plug syndrome has a similar presentation, but occurs in non-cystic fibrosis babies. Intestinal perforation does not complicate this condition.

(Choice 1) Intussusception can occur in patients with cystic fibrosis, but not this early. Its peak incidence is at 2 years of age, and manifests as abdominal pain and red currant jelly stools.

(Choice 4) Duodenal atresia can present with bilious vomiting in a newborn with an antenatal history of polyhydramnios, but the "double bubble" appearance on x-rays is usually present.

(Choice 5) Esophageal atresia is usually present in the setting of tracheoesophageal fistula. Although it is sometimes seen with polyhydramnios, it is an unlikely diagnosis in this case since bilious vomiting does not occur with esophageal atresia.

190. Question

1 points

Category: Pediatrics

A 9-month-old infant is brought to the emergency department with lethargy and tachypnea. He was healthy before developing fever and diarrhea four days ago. He has been taking some formula, but has had two to three episodes of diarrhea with each bottle. He has lost three pounds (1.4 kg) since his routine check-up two weeks ago. He has had one wet diaper in the past twenty four hours. On examination, his temperature is 102.5°F (39.1 °C), pulse is 200/min, respiratory rate is 42/min, and blood pressure is 70/45 mm Hg. He is lethargic with decreased tone and decreased deep tendon reflexes. His mucous membranes are dry. Cardiopulmonary exam reveals tachycardia and tachypnea. His abdominal exam is unremarkable. Capillary refill is four seconds. Laboratory results are shown below.

Chemistry panel: 165 mEq/L

Serum sodium: 4.5 mEq/L

Serum potassium: 108 mEq/L

Chloride Bicarbonate: 14 mEq/L

Blood urea nitrogen (BUN): 20 mg/dl

Serum creatinine: 0.8 mg/dl

Calcium: 10.0 mg/dl

Blood glucose: 98 mg/dl

Which of the following fluids should be used as a bolus in the resuscitation of this infant?

1. ☒ 0.9% saline ✓
2. ☐ 0.45% saline
3. ☐ 5% dextrose
4. ☐ 5% albumin
5. ☐ Packed red blood cells

INCORRECT ✗

The correct answer is 1.

The signs and symptoms of hypernatremia are mainly neurologic and include lethargy, altered mental status, irritability, and seizures. Hypernatremia can also cause muscle cramps, muscle weakness, and decreased deep tendon reflexes. Hypovolemic hypernatremia develops secondary to renal losses (e.g., diuretic use, glycosuria) or extrarenal losses (e.g., gastrointestinal upset, excessive sweating). Hypervolemic hypernatremia occurs due to exogenous sodium intake or mineralocorticoid excess (e.g., hyperaldosteronism). When treating a patient with hypernatremia, the sodium must be slowly returned to normal. In this infant with hypernatremia and dehydration, the initial goal is to stabilize him with fluid resuscitation as needed. When giving intravenous fluid boluses, only isotonic solutions such as normal saline or lactated Ringer's should be used.

(Choices 2 & 3) Half normal saline and 5% dextrose are hypotonic solutions. As such, they should never be used for initial resuscitation because they quickly exit the intravascular system and lower the sodium too rapidly. Precipitous drops in sodium levels can cause cerebral edema.

(Choice 4) Multiple studies have demonstrated that the expensive colloid solutions are no better than crystalloids at fluid resuscitation.

(Choice 5) Packed red blood cells may be appropriate in initial fluid resuscitation when bleeding is a major issue. Because of the limited availability of packed red blood cells on short notice, however, isotonic solutions remain the mainstay of initial fluid resuscitation (even in individuals with massive bleeding).

191. Question

1 points

Category: Pediatrics

A neonate is noted to have aniridia of the right eye on physical examination. He was born by spontaneous vaginal delivery after an uncomplicated full-term pregnancy. The remainder of the physical examination is normal. Which of the following is the most appropriate next step before the infant is released from the hospital?

1. ☒ An abdominal ultrasound ✓
2. ☐ An echocardiogram
3. ☐ A neurology consult
4. ☐ A rapid plasmin reagin (RPR) test
5. ☐ IV antibiotics

INCORRECT ✗

The correct answer is 1.

Aniridia describes the absence of an iris. It can be inherited as an autosomal dominant trait or it can occur sporadically. Approximately one third of sporadic cases are associated with Wilms tumor. Hemihypertrophy is also associated with Wilms tumor in some individuals. It is important to obtain an abdominal ultrasound to investigate for renal abnormalities or Wilms tumor in such cases.

(Choice 2) is not indicated.

(Choice 3) is not required because aniridia and Wilms tumor are not associated with acute nervous system disease. An ophthalmology consult would be more appropriate.

(Choice 4) is not warranted because syphilis is not suggested by the clinical findings or history. Neurosyphilis is associated with the Argyll-Robertson pupil, which irregularly shaped and nonreactive to light.

(Choice 5) are not indicated because aniridia is not associated with in utero or perinatal infection.

192. Question

1 points

Category: Pediatrics

A 3-year-old child is evaluated in the emergency department for a lump on his head, which his mother says appeared after he accidentally hit his head while jumping on the bed. According to the mother, the patient fell from the bed onto a non-carpeted wooden floor. The child did not lose consciousness, and did not have emesis, visual changes, or changes in behavior. Examination reveals a tense swelling over the right parietal area and no neurologic deficits. Which of the following is the most likely diagnosis?

1. ☐ Epidural hematoma
2. ☐ Subdural hematoma
3. ☐ Intraventricular hemorrhage
4. ☒ Subgaleal hematoma ✓
5. ☐ Lipoma

INCORRECT ✗

The correct answer is 4.

This patient has a subgaleal hematoma, which is a result of blood collecting sub adjacent to the galea. It is seen after minor injury and can be very alarming. It is not associated with loss of consciousness. The best treatment for this condition is to leave it alone. Aspirating the hematoma would only introduce infection, which could be disastrous. The parent should be reassured that no treatment is necessary.

(Choice 1) results from blood collecting within the cranial cavity, and is usually seen after a tear of the middle meningeal artery as it courses on the inner aspect of the temporal bone, in which a fracture line is seen. It is associated with a lucid interval and signs of raised intracranial pressure (i.e., headache, vomiting, loss of consciousness, enlarging pupil on the ipsilateral side, and hemiplegia on the contralateral side).

(Choice 2) results from intracranial hemorrhage within the subdural space. It may be acute or chronic. A fracture may be absent. The patient is comatose from the time of injury and has evidence of cerebral injury. The prognosis is poor.

(Choice 3) is associated with coma and may result from trauma. However, it is more commonly seen after hypertensive intracranial hemorrhage, in which the bleeding spreads to the ventricular system.

(Choice 5) is a benign soft-tissue tumor and does not develop suddenly.

193. Question

1 points

Category: Pediatrics

You are reviewing the vaccine schedule with a group of nursing students. You explain that the measles mumps-rubella vaccine (MMR) is a live attenuated vaccine that is generally given at 12–15 months, with a booster at 4–6 years. One of the students asks if there are any groups excluded from receiving the MMR vaccine. Your response is yes; all but one of the following groups should receive the MMR vaccine. Which one of the following groups should receive the vaccine?

- ☐ Pregnant women, persons with fever and upper respiratory infection, the immunocompromised, and those with allergy to neomycin causing contact dermatitis
- ☐ Pregnant women, patients with leukemia in remission, persons with moderate to severe fever, those with allergy to neomycin causing contact dermatitis
- ☐ Pregnant women, persons with no fever and upper respiratory infection, the immunocompromised, and those with allergy to neomycin causing contact dermatitis
- ☐ Pregnant women, patients positive for human immunodeficiency virus (HIV) not severely immunocompromised, persons with moderate to severe fever, and those with allergy to neomycin causing contact dermatitis
- ☒ Pregnant women, immunocompromised persons, persons with moderate to severe fever, and those with allergy to neomycin causing anaphylaxis ✓

INCORRECT ✗

The correct answer is 5.

Live vaccines, such as measles-mumps-rubella (MMR), should not be administered to pregnant woman, immunocompromised patients, persons with moderate to severe fever, and those with allergy to neomycin causing anaphylaxis.

(Choices 1 & 3) Persons with fever and upper respiratory infection (Choice 1) and persons with no fever and upper respiratory infections

(Choice 3) are not contraindications to the MMR vaccine. Likewise, persons with allergy to neomycin causing contact dermatitis (choices 1, 2, 3, & 4) should receive the MMR vaccine.

(Choice 2) Live-virus vaccines may be administered to persons with leukemia in remission who have not received chemotherapy in 3 months.

(Choice 5) However, the MMR vaccine should not be administered to persons with allergy to neomycin causing anaphylaxis.

(Choice 4) The not severely immunocompromised human immunodeficiency virus (HIV) positive patient should receive the MMR vaccine because of the increased risk associated with measles infection in this population, as well as the absence of serious adverse events after measles vaccination among this group.

194. Question

1 points

Category: Pediatrics

An 18-month-old male is brought to the hospital because of fever, dyspnea, and productive cough of two days duration. His mother reports that he just recovered from prolonged diarrhea due to Giardia infection. His past medical history is also significant for pneumonia and recurrent ear infections since 6 months of age. On physical examination, his temperature is 38.7 °C (101.7 °F), pulse is 140/min, and respirations are 40/min. Examination reveals a young child in mild respiratory distress and bronchial breath sounds in the right lower lung lobe. Which of the following is the most likely cause of his recurrent infections?

1. ☐ Impaired oxidative metabolism
2. ☐ Complement deficiency
3. ☐ Thymic hypoplasia
4. ☐ Adenosine deaminase deficiency
5. ☒ Abnormal B-lymphocyte maturation ✓

INCORRECT ✗

The correct answer is 5.

This infant has a history of Giardia and recurrent sinopulmonary infections since six months of age, suggesting a genetic defect in B-lymphocyte maturation. Genetic B-cell deficiencies present after 6 months of age because this is when the newborn's levels of maternal antibodies drop. Affected infants are predisposed to recurrent sinopulmonary infections with encapsulated organisms like H. influenzae and S. pneumoniae because immune destruction of encapsulated organisms depends upon the humoral immune response. Deficiency in IgA, which inhabits the GI tract, is the specific deficiency that predisposes to Giardia infection.

(Choice 1) Chronic granulomatous disease (CGO) is a condition of impaired oxidative metabolism. In CGO, a defect in the NADPH-oxidase system of phagocytic cells results in defective intracellular killing. Patients are therefore prone to abscesses secondary to catalase-producing organisms like Aspergillus and Staphylococcus.

(Choice 2) Gonococcal and meningococcal infections are common in patients with complement deficiency. S. pneumoniae and H. influenza infections do occur in this patient population as well, but infections with Giardia are not typically seen.

(Choice 3) Thymic hypoplasia is one element of DiGeorge syndrome. The resultant T-cell deficiency puts patients at high risk for viral and fungal infections.

(Choice 4) Adenosine deaminase deficiency is commonly the genetic defect underlying severe combined immunodeficiency (SCID). Patients with SCID have a deficiency of both B- and T-cells. The T-cell deficiency predisposes to viral and fungal infections and the B-cell deficiency predisposes to infection by bacteria.

195. Question

1 points

Category: Pediatrics

A 3-month-old girl is brought to the pediatrician for a scheduled visit. She has been meeting all development milestones but has been vomiting after each feeding. The infant weighed 3 kg (6 lb 10 oz) at birth and now weighs 6 kg (13 lb 3 oz). She does not have diarrhea and is afebrile. The remainder of the physical examination is unremarkable. Which of the following is the most likely cause of this patient's vomiting?

1. ☐ Adrenogenital syndrome
2. ☐ Child abuse
3. ☐ Inborn error of metabolism
4. ☒ Overfeeding ✓
5. ☐ Pyloric stenosis

INCORRECT ✖

The correct answer is 4.

Usually, infants gain an ounce/ day during the first 4 months of life. Those who gain more than this and are spitting up are probably being overfed.

(Choice 1) Adrenogenital syndrome, or congenital adrenal hyperplasia, presents in the first week of life with metabolic abnormalities and results in poor weight gain.

(Choice 2) Children being abused would probably not be overfed.

(Choice 3) present in the first weeks of life and generally lead to poor weight gain.

(Choice 5) typically presents with projectile vomiting at 2 weeks of age. It is more common in boys.

196. Question**1 points****Category: Pediatrics**

A 12-year-old girl is taken to a pediatrician complaining of a sore mouth. On questioning, the child states that she has been feeling poorly, with fatigue and weakness. She began menstruating briefly and then stopped. Physical examination is notable for focal white crusting of the oral cavity; biopsy of one of these areas later shows candidiasis. Laboratory studies show the following:

Sodium: 127 mEq/L**Potassium:** 5.3 mEq/L**Bicarbonate:** 24 mEq/L**Calcium:** 7.5 mEq/dL**Phosphorus:** 5.5 mg/dL**Glucose:** 87 mg/dL

Which of the following is the most likely diagnosis?

1. ☐ Multiple endocrine neoplasia, type I
2. ☐ Multiple endocrine neoplasia, type II A
3. ☒ Polyglandular deficiency syndrome, type I
4. ☐ Polyglandular deficiency syndrome, type II
5. ☐ Polyglandular deficiency syndrome, type III

INCORRECT ✖

The correct answer is 3.

The polyglandular deficiency syndromes are autoimmune and are characterized by concurrent subnormal function of several endocrine glands. This child has the type I form, which can occur in children or younger adults, with peak incidence at 12 years. Polyglandular deficiency syndrome, type I, frequently has chronic mucocutaneous candidiasis, hypoparathyroidism (as evidenced by hypocalcemia and hyperphosphatemia), and Addison disease (adrenocortical failure, as indicated by hyponatremia and low bicarbonate). Other features encountered with some frequency in this condition are gonadal failure, alopecia, malabsorption, and, much less commonly, thyroid disease, pernicious anemia, diabetes mellitus, vitiligo, and chronic active hepatitis.

(Choice 1) is characterized by parathyroid adenomas with hypercalcemia, pancreatic islet cell tumors, and pituitary adenomas.

(Choice 2) is characterized by medullary carcinoma of the thyroid (which might produce hypocalcemia), Pheochromocytomas (which would produce hypertension), and sometimes parathyroid adenomas (which might produce hypercalcemia).

(Choice 4) occurs in adults and is commonly characterized by adrenocortical insufficiency, thyroid disease, and diabetes mellitus.

(Choice 5) occurs in adults. It is similar to type II but specifically lacks adrenocortical insufficiency.

197. Question**1 points****Category: Pediatrics**

A 28-year-old Caucasian female comes to the office due to fever for the past 3 days. She also complains of pain and swelling in her right breast for the past 4 days. She exclusively breastfeeds her 3-month-old baby. On examination, she is febrile. Her right breast is engorged, red and tender, without any area of fluctuation. She wants to know if she can continue to breastfeed. Which of the following is a contraindication to breastfeeding?

1. ☐ Mastitis
2. ☒ Acquired immune deficiency syndrome ✓
3. ☐ Maternal rubella infection
4. ☐ Breast milk jaundice
5. ☐ Hemolytic disease of the newborn

INCORRECT ✗**The correct answer is 2.**

Although breastfeeding is generally encouraged due to its many advantages, physicians should always watch out for situations in which breastfeeding may do more harm than good. Some contraindications to breastfeeding include certain active maternal infections (i.e., HIV, tuberculosis, herpes simplex lesions on or near the nipple, malaria, sepsis, typhoid fever), eclampsia, nephritis, substance abuse, and breast cancer. Transmission of HIV by breastfeeding is well documented; therefore, the presence of maternal HIV infection is a contraindication to breastfeeding.

(Choice 1) Mastitis is not a contraindication to breastfeeding. Some physicians actually encourage breastfeeding since this prevents engorgement, along with other subsequent symptoms such as fissures and cracking of the nipple. The woman in this case most likely has mastitis.

(Choice 3) Maternal rubella infection is not a contraindication to breastfeeding since there is currently insufficient evidence regarding the associated risks.

(Choice 4) Breast milk jaundice is not a contraindication to breastfeeding. Mothers of such infants are instructed to switch their babies to formula-feeding for 1-2 days. This allows the increased indirect bilirubin levels of the infant to return to normal values. After 2 days, the mother resumes breastfeeding, and further hyperbilirubinemia usually does not recur.

(Choice 5) Hemolytic disease of the newborn (erythroblastosis fetalis) is not a contraindication to breastfeeding because antibodies in the mother's milk are inactivated in the intestinal tract and do not contribute to further hemolysis of the infant's RBCs.

198. Question

1 points

Category: Pediatrics

A 2-year-old boy is referred to your office for the evaluation of a white eye reflex. His mother suspects a hearing problem because he does not respond when she calls out his name. The physical examination reveals a continuous, machinery murmur over the right 2nd intercostal space and purple lesions on his arms and chest. The initial investigations reveal thrombocytopenia. What is the most likely diagnosis?

1. ☐ Sturge Weber syndrome
2. ☐ Retinoblastoma
3. ☒ Congenital rubella infection ✓
4. ☐ Congenital CMV infection
5. ☐ Congenital Toxoplasma infection

INCORRECT ✗

The correct answer is 3.

The child most likely has congenital rubella infection, which is transmitted via the placenta. The causative organism is Toga virus. Maternal infection manifests after an incubation period of 14 to 21 days with mild symptoms such as rash, arthralgias and generalized lymphadenopathy. Since rubella infection confers permanent immunity, only primary infection in the pregnant woman carries the risk of fetal disease. The severity of fetal disease depends on the time of pregnancy when the transmission took place. Infections of early pregnancy are the most severe, and the associated adverse outcomes include spontaneous abortion and congenital rubella syndrome (CRS). If transmission occurs in the first 4 weeks of pregnancy, the risk of developing CRS is 50%. This risk drops to 1 % if transmission occurs in the third trimester. The features of CRS may include IUGR, deafness, cardiac malformations (e.g., patent ductus arteriosus, atrial septal defects), microphthalmia, cataract, retinopathy, hepatosplenomegaly, thrombocytopenia, and CNS involvement. Chronic infection may result in growth retardation, radiolucent bone disease, jaundice, hepatosplenomegaly, thrombocytopenia and purple skin lesions ("blueberry muffin spots").

Maternal infection is confirmed by the presence of IgM to rubella virus, or a fourfold increase in IgG on two serum samples obtained two weeks apart. In the infant, the diagnosis is confirmed by the presence of IgM or persistence of IgG beyond the age of 6 months. Prevention of congenital rubella is primarily achieved by administration of rubella vaccine to all females of childbearing age. If the immunologic status of a pregnant woman is unknown, rubella titers should be obtained in the first trimester. Immunization should not be performed in pregnancy because of a theoretical risk for the fetus, and such women should be advised to avoid anyone with possible rubella infection

(Choice 1) Sturge-Weber syndrome is a neurocutaneous disease characterized by a port-wine stain on the trigeminal nerve distribution, mental retardation, seizures and glaucoma. The skin lesions of the above patient (purple and located in multiple areas of the body) are not characteristic of Sturge-Weber syndrome.

(Choice 2) Retinoblastoma may present with a unilateral or bilateral white light reflex with or without exophthalmos; however, it is not accompanied by deafness or cardiac anomalies, as in CRS.

(Choices 4 & 5) Congenital CMV and Toxoplasma are similar in many ways. They both manifest with hydrocephalus, cerebral calcification, chorioretinitis, microcephaly and microphthalmia. Although rubella may also present with such symptoms, the presence of specific symptoms such as deafness, cardiac anomalies and blueberry muffin spots clarify the correct diagnosis.

199. Question

1 points

Category: Pediatrics

A 3-year-old child with sickle cell disease presents to the emergency department with a temperature of 40°C (104°F). The mother states that the child had been in his usual state of relatively good health until 4 hours ago. Starting at that time, the mother states that the child became progressively listless. Upon physical examination, the physician notes that the child has nuchal rigidity. The patient has no known ill contacts and takes no medication. Immunizations are up to date. A spinal tap would most likely reveal which of the following pathogens?

1. ☒ Streptococcus pneumoniae ✓
2. ☐ Group B streptococcus
3. ☐ Escherichia coli
4. ☐ Listeria monocytogenes
5. ☐ Group A β -hemolytic streptococcus

INCORRECT ✗

The correct answer is 1.

Patients with sickle cell disease are at increased risk for developing severe pneumococcal disease because of impaired immunologic response to Streptococcus pneumoniae. Patients with sickle cell disease should be immunized with the polyvalent pneumococcal vaccine in addition to all other routine immunizations of childhood. The polyvalent pneumococcal vaccine should be administered to asplenic children 2 years of age and older. The pneumococcal vaccine may not be as effective in children under 5 years old as it is in older individuals. Even if patients with sickle cell disease are given prophylactic antibiotics, there is still a chance that they will develop pneumococcal meningitis before 5 years of age.

(Choices 2,3,4 & 5) Escherichia coli, Listeria monocytogenes, and group B streptococcus are more common pathogens in newborns. The most common sites of infection for group A β -hemolytic streptococci are the upper respiratory tract, skin, soft tissues, and blood.

200. Question

1 points

Category: Pediatrics

A 45-year-old woman with polyhydramnios delivers a full-term male infant with a birth weight of 6 lb 12 oz (3.06 kg). The infant's physical examination is pertinent for hypotonia, upward and slanted palpebral fissures and epicanthic folds, a simian crease, and a short fifth finger. A few hours after birth, the infant begins to vomit bile-stained fluid. On physical examination, the abdomen is not distended. Radiographs of the chest and abdomen were obtained, and the results revealed a "double-bubble" sign. Which of the following is the most likely diagnosis?

1. ☐ Congenital pyloric stenosis
2. ☒ Duodenal atresia ✓
3. ☐ An intussusception
4. ☐ A diaphragmatic hernia
5. ☐ A perforated viscus

INCORRECT ✗

The correct answer is 2.

The patient in this question has trisomy 21, or Down syndrome, which is characterized by hypotonia, a simian crease, a short fifth finger, and upward and slanted palpebral fissures and epicanthic folds. These patients will also have differing degrees of mental and growth retardation. Duodenal atresia is often associated with Down syndrome and appears in the first day of life as bilious vomiting after feedings without abdominal distention. Plain abdominal radiographs typically show the "double-bubble" sign, caused by a distended and gas-filled stomach and proximal duodenum. The initial treatment for patients with duodenal atresia is nasogastric or orogastric decompression and intravenous fluid replacement. The definitive treatment is duodenojejunostomy.

(Choice 1) appears at 2–3 weeks of age with non bilious vomiting.

(Choice 3) Bile-stained emesis may be seen in the later phase of intussusception; however, it is rare under 3 months of age. It is associated with colicky abdominal pain, a sausage-shaped mass, and currant-jelly stool.

(Choice 4) If a diaphragmatic hernia presents at birth, the infant usually has a scaphoid abdomen and respiratory distress due to herniation of the abdominal contents into the chest that compresses the lungs. Bowel sounds can be auscultated in the chest, and the presence of abdominal contents in the thorax can be visualized on chest roentgenogram.

(Choice 5) will cause free air in the abdomen that is evident on plain abdominal radiographs as air under the diaphragm.

201. Question

1 points

Category: Pediatrics

A 4-month-old infant is brought to the pediatrician by his mother because he has not been feeding well. The patient has been in his usual state of good health until 1 day earlier. The patient has not had fever and does not attend day care. There are no ill contacts, pets, or recent travel. Physical examination of the mouth reveals curd like plaques on the tongue and buccal mucosa that do not scrape off easily. Which one of the following is the bodily area most likely to also be affected?

1. ☐ Eyes
2. ☐ Ears
3. ☐ Nose
4. ☐ Scalp
5. ☒ Perineum

INCORRECT ❌

The correct answer is 5.

Candida species are common causes of oral thrush and diaper dermatitis in infants. Thrush consists of white, curd-like plaques on the tongue, gums, and buccal mucosa that are difficult to remove. The skin manifestations include a beefy-red maculopapular rash that may coalesce and form satellite lesions. Intertriginous areas, especially in the diaper area (perineum), also are usually involved.

(Choices 1,2,3 & 4) Candida infections of the eyes, ears, nose, and scalp are not usually associated with thrush. Disseminated Candida infection is most commonly associated with an immunocompromised host.

202. Question

1 points

Category: Pediatrics

A 26-day-old infant is brought to the emergency department with a temperature of 39.0°C (102.2°F). The mother is a 25-year-old gravida 2, para 2 who had good prenatal care. Samples of the cerebrospinal fluid (CSF), blood, and urine are sent for culture and analysis. In addition, prophylactic antibiotics are started. It is of paramount importance that the antibiotic treatment provides protection against which of the following pathogens?

- ☐ Escherichia coli, Streptococcus pneumoniae, and Listeria monocytogenes
- ☐ Streptococcus pneumoniae, Neisseria meningitidis, and Listeria monocytogenes
- ☒ Streptococcus agalactiae, Escherichia coli, and Listeria monocytogenes ✓
- ☐ Streptococcus agalactiae, Escherichia coli, and Neisseria meningitidis
- ☐ Neisseria meningitidis, Streptococcus pneumoniae, and Streptococcus agalactiae

INCORRECT ✗

The correct answer is 3.

The infectious disease that the physician would be most concerned about is meningitis. The treatment must be designed to prevent or, if already fulminating, cure this disease. Streptococcus agalactiae (group B streptococcus) is a common inhabitant of the maternal genitourinary tract. It is acquired by newborns via vertical transmission, and it is the most common cause of neonatal meningitis, followed by Escherichia coli and Listeria monocytogenes. Early-onset disease is associated with sepsis; late onset is associated with meningitis. Consequently, S. agalactiae, E. coli, and L. monocytogenes is the correct answer.

(Choices 1,2,4 & 5) Streptococcus pneumoniae and Neisseria meningitidis are more commonly seen in older age groups.

203. Question

1 points

Category: Pediatrics

You are called to examine a one-day-old male neonate who gradually developed cyanosis over the past few hours. The infant was delivered vaginally at full term, assisted with forceps, and weighed 9 lbs. The Apgar scores were 7 and 8 at 1 and 5 minutes, respectively. Oxygen administration by mask does not relieve the cyanosis. Further examination reveals tachypnea, subcostal retractions, a normal S₁, single and loud S₂, and no murmur. Which of the following is the most probable cause of the infant's cyanosis?

1. ☒ Transposition of great vessels ✓
2. ☐ Atrial septal defect
3. ☐ Coarctation of aorta
4. ☐ Ventricular septal defect
5. ☐ Patent ductus arteriosus

INCORRECT ✗

The correct answer is 1.

Transposition of great vessels is a cyanotic heart disease which presents with cyanosis in the first 24 hours of life. It is commonly seen in infants of diabetic mothers and in males. The aorta arises from the right ventricle and the pulmonary artery from the left ventricle. As a result, deoxygenated blood coming from the body goes to the right atrium and the right ventricle, and is cycled back to the body through the aorta. Oxygenated blood from the lungs is returned to the lungs by the left side of the heart through the pulmonary artery.

During intrauterine life, the foramen ovale and ductus arteriosus provide mixing of the deoxygenated and oxygenated blood, resulting in an almost normal oxygenation of the fetal circulation; however, after birth, the foramen ovale and ductus arteriosus begin to close, leading to decreased mixing and poor oxygenation. The neonate then presents with cyanosis and tachypnea within the first few hours of life. X-ray of the chest may be normal initially, but may show some evidence of increased pulmonary blood flow after 1 to 2 weeks. Echocardiography confirms the diagnosis. Maintaining the patency of the ductus arteriosus is important for survival. Surgical treatment is definitive.

(Choices 2, 3, 4 & 5) Atrial septal defect, coarctation of aorta, ventricular septal defect, patent ductus arteriosus and dextrocardia are all acyanotic heart diseases.

204. Question

1 points

Category: Pediatrics

A 13-year-old basketball player presents to the pediatrician with a limp and right knee pain, which he states to have had for the last week. He tells the pediatrician that the basketball season has just begun and the training is physically demanding. The patient thinks that he may have pushed himself too hard during workouts. He reports that running, jumping, or climbing stairs exacerbates his knee pain. On physical examination, the tibial tuberosity is enlarged. However, there is no synovial effusion or thickening of the knee joint. Which one of the following is the most likely diagnosis?

1. ☐ Legg-Calve-Perthes disease
2. ☐ Slipped capital femoral epiphysis
3. ☒ Osgood-Schlatter disease ✓
4. ☐ Septic arthritis of the knee
5. ☐ Idiopathic adolescent anterior knee pain syndrome

INCORRECT ✖

The correct answer is 3.

Osgood-Schlatter disease is an overuse injury that is more common in physically active boys around puberty. It is characterized by pain and swelling of the tibial tubercle. The treatment is focused on reduction of the activity. Anti-inflammatory medications are usually not helpful. The condition is usually self-limiting and resolves in 12–24 months.

(Choice 1) presents with a gradual limp, stiffness, and pain in the groin, hip, thigh, or knee. This avascular necrosis of the femoral head usually occurs between the ages of 2 and 12 years (mean 7 years).

(Choice 2) typically occurs in adolescents who are overweight and have delayed skeletal maturity, or in those who are tall and thin and have had a recent growth spurt. The chronic condition is associated with endocrine abnormalities such as hypothyroidism.

(Choice 4) is an infection within a joint space causing pain and limitation of motion. Analysis of the joint fluid is mandatory for diagnosis.

(Choice 5) Patients with idiopathic adolescent anterior knee pain syndrome, formally known as chondromalacia patellae, develop knee pain of unknown cause in early adolescence. Strenuous physical activity, such as running, may cause knee pain in these adolescents. However, there are no other associated findings.

205. Question

1 points

Category: Pediatrics

A pediatrician is called to the delivery room because a full-term infant has developed cyanosis and respiratory distress immediately after birth. A brief examination of the infant reveals cyanosis on room air not completely relieved by oxygen administered by mask, subcostal and intercostal retractions, absent air entry on the left with audible bowel sounds in the left chest, and poor air entry on the right chest. The heart is best heard in the right hemithorax; the abdomen is flat without organomegaly. Which of the following is the most likely diagnosis?

1. ☒ Congenital diaphragmatic hernia ✓
2. ☐ Hyaline membrane disease
3. ☐ Meconium aspiration
4. ☐ Pneumonia
5. ☐ Tracheoesophageal fistula

INCORRECT ✗

The correct answer is 1.

Typically, the abdominal contents herniate into the left hemithorax. This causes displacement of the heart into the right hemithorax and pulmonary hypoplasia. If gas has entered the intestinal tract during resuscitation, bowel sounds may be heard in the chest. The displacement of the abdominal contents renders the abdomen flat. In cases of severe pulmonary hypoplasia, administration of oxygen results in only a poor improvement in oxygenation.

(Choice 2) is uncommon, if not rare, in full-term infants. Occasionally this form of respiratory distress may occur in infants of diabetic women, but this history is absent in this case.

(Choice 3) should be accompanied by a history of fetal stress, meconium-stained amniotic fluid, or meconium-staining of the skin and cord. Physical findings are symmetric, there is no displacement of the heart, and breath sounds are similar bilaterally.

(Choice 4) in the neonate is usually due to either group B β -hemolytic streptococci or Escherichia coli, accompanied by a history of maternal fever or prolonged rupture of the amniotic membranes. Breath sounds are symmetrically diminished.

(Choice 5) usually presents with a history of polyhydramnios, cyanosis with feeding, and increased oropharyngeal secretions. It is unusual for this condition to present so soon after birth with severe respiratory distress.

206. Question

1 points

Category: Pediatrics

A 5-year-old girl with severe mental retardation is brought to the pediatrician for immunizations. According to the history, the patient appeared to be developing normally until 18 months of age, when she acquired dementia and her head circumference plateaued, resulting in microcephaly. She wrings her hands, and sighs. She is noted to have ataxia and marked loss of gross motor skills. In addition, she has loss of language milestones. Which of the following is the most likely diagnosis?

1. ☐ Autism
2. ☐ William syndrome
3. ☒ Rett syndrome ✓
4. ☐ Turner syndrome
5. ☐ Russell-Silver syndrome

INCORRECT ✗

The correct answer is 3.

Rett syndrome occurs only in females. Girls with Rett syndrome appear to be developing normally until 6–18 months of age, when they manifest dementia, develop microcephaly, and lose purposeful movements of their hands (i.e., hand-wringing). They may also have ataxia, seizures, scoliosis, and loss of gross motor skills. They may have sighing respirations associated with apnea and cyanosis.

(Choice 1) may manifest with great variability in severity and range of symptoms. The usual presenting complaint is regression of language and play without loss of motor skills.

(Choices 2 & 4) Characterizations of William syndrome include a depressed nasal bridge, epicanthal folds, prominent thick lips with open mouth, and short stature. Short stature, webbed posterior neck, broad chest with widely spaced nipples, and an increased carrying angle of the arms characterize Turner syndrome.

(Choice 5) Key features of Russell-Silver syndrome are short stature, triangular facies with down-turned corners of the mouth, clinodactyly, and a head that appears disproportionately large.

207. Question

1 points

Category: Pediatrics

A 7-year-old boy is brought to the office by his parents because his grades are deteriorating. According to the mother, the child is always preoccupied and prefers to play with 'part of the toy rather than the entire toy.' The child's IQ is 55. On examination, the child has a very poor attention span. He has a prominent jaw, long ears and large testicles. What is the most likely cause for this child's mental retardation?

1. ☐ Trisomy 21
2. ☐ Maternal alcohol use
3. ☒ Increased number of CGG trinucleotide repeats ✓
4. ☐ Attention-deficit hyperactivity disorder
5. ☐ Autistic disorder

INCORRECT ✗

The correct answer is 3.

Affected males of Fragile X syndrome may have a characteristic appearance (large head, long face, prominent forehead and chin, protruding ears), joint laxity, and large testes. Behavioral abnormalities, including hyperactivity, short attention span, and autism, are common. The syndrome results from a full mutation in the FMR 1 gene caused by an increased number of CGG trinucleotide repeats accompanied by aberrant methylation of the FMR 1 gene.

(Choice 1) Down's syndrome is characterized by mental retardation, microcephaly, flat occiput, Simian crease, endocardial cushion defects, prominent tongue, epicanthal folds with slanting eyes, and Brush field's spots on the periphery of the iris.

(Choice 2) Fetal alcohol syndrome is characterized by mild to moderate mental retardation, hypo-plastic maxilla, long philtrum and microcephaly.

(Choices 4 & 5) Attention deficit hyperactivity disorder and autistic disorder may be presenting features of Fragile X syndrome, but by themselves do not explain the other clinical findings in this child.

208. Question

1 points

Category: Pediatrics

A blood type B infant born to a blood type O mother has clinically significant fetal-maternal blood group incompatibility with mild anemia and a weakly positive Coombs test. The infant develops jaundice a few hours after birth, with a bilirubin (measured at 12 hours after birth) of 12 mg/dL (predominately unconjugated) compared with 3.5 mg/dL in cord blood. The physician is concerned that the rising bilirubin levels will damage the infant's nervous system. Which of the following sites is most vulnerable to this injury?

1. ☒ Basal ganglia ✓
2. ☐ Cerebellum
3. ☐ Cerebral cortex
4. ☐ Peripheral nerve
5. ☐ Spinal cord

INCORRECT ✗

The correct answer is 1.

The term kernicterus is used to denote damage to the brain by high bilirubin levels in the perinatal period. The fetal liver is less efficient at conjugating bilirubin compared with the adult liver, and neonates are consequently prone to develop hyperbilirubinemia, particularly unconjugated hyperbilirubinemia. In most cases, there are predisposing conditions, such as hemolytic anemia, polycythemia, prematurity, maternal diabetes, congenital liver or biliary tract disease, sepsis, or intrauterine infections. In contrast to what you might expect, the bilirubin is not uniformly poisonous to the nervous system, but instead selectively accumulates in, damages, and stains the basal ganglia and the brain stem nuclei. Early symptoms may include lethargy, poor feeding, and vomiting. Severe kernicterus can cause opisthotonos, oculogyric crisis, seizures, and even death.

(Choices 2,3,4 & 5) The cerebellum, cerebral cortex, peripheral nerve, and spinal cord are all much less sensitive to damage by high bilirubin levels than are the basal ganglia and brain stem nuclei.

209. Question

1 points

Category: Pediatrics

A 3-year-old boy is brought to the office by his 27-year-old white mother for the evaluation of recurrent bone fractures. His first fracture was that of the femur, and occurred when he was 6 months old. He had a fracture of the wrist 4 months ago. His mother also has a history of multiple fractures since childhood. She lost all her teeth at a very early age and is complaining of deafness. Her husband has a history of severe alcohol abuse. On examination, both mother and son have blue sclerae. What is the most likely involved disease process?

1. ☒ Mutations in type 1 collagen ✓
2. ☐ Mutations in fibrillin 1 gene
3. ☐ Child abuse
4. ☐ Vitamin-D deficiency
5. ☐ Congenital syphilis

INCORRECT ✗

The correct answer is 1.

The above child is most likely suffering from osteogenesis imperfecta, which is an inherited connective tissue disorder caused by a mutation in the genes coding for type I collagen. Since type I collagen is an important structural protein that is present in the skin, sclera, bone, tendon and ligament, patients with this disorder present with multiple recurrent fractures, blue sclera, hearing loss, joint laxity, short stature, and scoliosis.

(Choice 2) Marfan syndrome is one of the most common connective tissue disorders, and is caused by mutation of the fibrillin-1 gene. Its mode of inheritance is autosomal dominant. The common features of this disorder include skeletal manifestations (e.g., arachnodactyly, hypermobility of joints), ectopia lentis, and aortic root dilatation.

(Choice 3) Child abuse suspected in the presence of any of the following: retinal hemorrhages, burns, bruising, fractures or abrasions. This is a good differential diagnosis for the given case; however, the features of this patient (e.g., fractures, blue sclerae, family history of fractures and hearing loss) are very typical of osteogenesis imperfecta.

(Choice 4) Children with rickets have slow growth and skeletal deformities. Blue sclerae and hearing loss are not the features of rickets.

(Choice 5) Early manifestations of congenital syphilis include jaundice, rash, lymphadenopathy, rhinitis and hepatosplenomegaly. Late manifestations occur around two year of age; these include saber shins, keratitis, Hutchinson's teeth, saddle-nose deformity and deafness.

The above child has typical features of osteogenesis imperfecta, not congenital syphilis.

210. Question

1 points

Category: Pediatrics

A 3-year-old presents with a history of daily fever with temperature spikes of 39°C for the past 2 weeks. The parents state that the fever is followed by an evanescent rash over the trunk and extremities, as illustrated above. In addition, they say that the patient has arthritis-like pains. The patient also has hepatosplenomegaly, lymphadenopathy, and a pericardial effusion. The erythrocyte sedimentation rate (ESR) is elevated, and there is an absolute neutrophilic leukocytosis. Which of the following is the most likely diagnosis?

1. ☐ Acute rheumatic fever
2. ☐ Systemic lupus erythematosus (SLE)
3. ☒ Juvenile rheumatoid arthritis (JRA) ✓
4. ☐ Lyme disease
5. ☐ Ankylosing spondylitis

INCORRECT ✗

The correct answer is 3.

Juvenile rheumatoid arthritis (JRA) is an autoimmune inflammatory disease that causes chronic synovitis as well as other extraarticular manifestations. This arthritis is divided into subgroups on the basis of presentation and laboratory findings. Systemic JRA occurs in approximately one-fifth of patients and is associated with high fevers, rash, hepatosplenomegaly, pleural and pericardial effusions, and anemia of chronic disease. The erythrocyte sedimentation rate (ESR) is elevated during active disease.

(Choice 1) can be differentiated from JRA by the evidence of carditis and its transient migratory arthritis.

(Choice 2) usually has milder joint manifestations and a characteristic malar rash.

(Choice 4) should be considered in the differential diagnosis. A careful history and physical examination, searching for tick bites and characteristic rash of erythema chronicum migrans, should be performed.

(Choice 5) can be mistaken for JRA until spine involvement is seen. Its onset is later in life, and it is much more common in men.

211. Question

1 points

Category: Pediatrics

Within 8 hours afterbirth, an infant has «excessive salivation."Physical examination reveals that she has an imperforate anus, with a small fistula to the vagina. A small, soft nasogastric tube is inserted, and the infant is taken to x-ray. The film shows the tube coiled back on itself in the upper chest, and a normal gas pattern in the gastrointestinal tract. There are no apparent abnormalities of the radius or the vertebral bodies. Which of the following is the most appropriate next step in management?

1. ☒ Renal sonogram and echocardiogram ✓
2. ☐ Barium swallow
3. ☐ Placement of a gastrostomy tube
4. ☐ Diverting colostomy
5. ☐ Surgical repair of esophageal atresia

INCORRECT ✗

The correct answer is 1.

This infant already has evidence of imperforate anus and tracheoesophageal fistula with the most common configuration (blind esophageal end at the top, and fistula to the tracheobronchial tree at the bottom that is how gas got into the gastrointestinal tract). Those two anomalies are part of the VACTERL constellation (vertebral, anal, cardiac, tracheoesophageal, renal, and limb abnormalities). The x-ray already ruled out vertebral and radial anomalies, but we still have to rule out cardiac and renal anomalies. For that reason, the infant needs the renal sonogram and the echocardiogram. No surgical repair should be planned until we know the full extent of the problem.

(Choice 2) is not the first thing we need. If further definition of the esophageal anomaly is desired, a more gentle medium (Gastrografin) should be used to avoid aspiration of barium.

(Choice 3) Gastrostomy may very well be needed if the full extent of the anomalies delays repair of the tracheoesophageal fistula. Reflux of gastric contents into the tracheobronchial tree will produce chemical damage. However, it does not take that long to do a sonogram of the kidney and an echocardiogram, and if they are negative, repair of the tracheoesophageal anomaly may be undertaken at once.

(Choice 4) Colostomy is not urgent, because there is a fistula from the end of the gastrointestinal tract to the vagina. A decision as to the type of repair possible now, or to be deferred until later can be made at leisure.

(Choice 5) Surgical repair has to wait until we know the full extent of the anomalies. The idea that diagnosis must precede therapy is well illustrated in this case.

212. Question

1 point

Category: Pediatrics

A 3-year-old child is taken to a pediatrician because he develops burning pain, erythema, and swelling minutes after being exposed to the sun. Physical examination demonstrates erythema with swelling of the hands and arms. The skin is thickened on the backs of the hands but does not show blistering or scarring. Which of the following is the most likely diagnosis?

1. ☐ Acute intermittent porphyria
2. ☒ Erythropoietic protoporphyria ✓
3. ☐ Hepatoerythropoietic porphyria
4. ☐ Porphyria cutanea tarda
5. ☐ Variegata porphyria

INCORRECT ✗

The correct answer is 2.

Erythropoietic protoporphyria is the most common form of erythropoietic porphyria. The clinical presentation illustrated is typical. Patients may also present with liver function test abnormalities. Protoporphyrin concentrations in plasma and red cells are markedly increased in these cases, whereas urine Porphyrins are not. Lead poisoning can also raise red cell protoporphyrin concentrations, so this may need to be excluded in a young child. Beta carotene is particularly effective in treating this type of porphyria, but it must be taken in amounts large enough to cause slight yellowing of the skin.

(Choice 1) usually presents with severe abdominal pain.

(Choice 3) is a very rare condition that can have a presentation similar to that in the question stem.

(Choice 4) causes chronic skin blistering.

(Choice 5) can present with either abdominal pain or chronic skin blistering.

213. Question

1 points

Category: Pediatrics

A first-year pediatric resident is called to the newborn nursery to evaluate a 2-day-old who has passed a bloody stool. The patient was born by normal spontaneous vaginal delivery to a 34-year-old primigravida. The birth weight was 8 lb 3 oz (3.7 kg), and Apgar scores were 8 at 1 minute and 9 at 5 minutes. The mother had good prenatal care throughout the entire pregnancy. She denied using tobacco, alcohol, or drugs during her pregnancy. The patient has been sleeping, feeding, defecating, and urinating well since birth. Vital signs are stable, and the rest of the physical examination findings are within normal limits. Which of the following tests would best determine the cause of the bloody stool?

1. ☒ Apt test on the blood in the stool ✓
2. ☐ Hemoglobin electrophoresis of the blood from the stool
3. ☐ Barium enema
4. ☐ Clinitest examination of the stool
5. ☐ Methylene blue stain of the stool

INCORRECT ✗

The correct answer is 1.

The most common cause of blood in the stool of a newborn is swallowed maternal blood. The Apt test helps distinguish adult hemoglobin (Hb) from fetal Hb. It is easier and less expensive than electrophoresis (**Choice 2**). If adult Hb is detected, the blood swallowed is maternal blood. If fetal Hb is found, a search begins for the cause of the bleeding.

(**Choice 3**) is both diagnostic and therapeutic for intussusception, which is rare before 3 months of age.

(**Choice 4**) is used to quantify the urinary level of reducing sugars.

(**Choice 5**) The presence of leukocytes in the stool is determined by the methylene blue stain.

214. Question

1 points

Category: Pediatrics

An obese 18-year-old presents at an emergency clinic in labor and asks for help in delivering her first child. The triage nurse takes a history and determines that the patient has had no prenatal care and cannot recall when her last menstrual cycle was, as she has always had irregular cycles. She further reveals that she first realized that she might be pregnant about 4 months ago; she denies the usage of drugs, alcohol, or tobacco and also denies having any sexually transmitted diseases. The baby was quickly delivered with no untoward incidences. At delivery, the infant is noted to be large for gestational age and has cracked, peeling skin. The infant's fingernails are long and its fingers are stained green. There is absence of lanugo. Which of the following is the most likely gestational age of this infant?

1. ☐ 34 weeks
2. ☐ 36 weeks
3. ☐ 38 weeks
4. ☐ 40 weeks
5. ☒ 42 weeks ✓

INCORRECT ✗

The correct answer is 5.

Infants born before 37 weeks' gestation (**Choices 1 & 2**) are preterm. Preterm infants may have lanugo hair, smooth, pink skin with visible veins, faint plantar creases, and ears that are soft with slow recoil. Term infants are defined as those who are born between 37 and 42 weeks' gestation (**Choices 3 & 4**). Term infants have a small amount of lanugo and some bald areas; smooth, slightly thickened skin; ears that have good recoil; and indentations over more than the anterior third of the plantar area. Post term babies are born after 42 weeks' gestation. They tend to be large for gestational age and hyper alert. Lanugo is absent, and the skin is cracked and peeling. Fingernails are long. They are at increased risk for meconium aspiration.

215. Question

1 points

Category: Pediatrics

A 12-year-old boy presents with an intensely pruritic rash for 3 days. He just went on a camping trip, during which he wore only short-sleeve shirts and short pants. His temperature is 37.6 C (99.7 F), blood pressure is 96/62 mm Hg, pulse is 65/min, and respirations are 12/min. There are numerous erythematous papules and vesicles on both arms and legs. Most of them are in a linear array. Which of the following is the most appropriate pharmacotherapy?

1. ☐ Oral cephalalexin
2. ☒ Oral prednisone ✓
3. ☐ Topical diphenhydramine
4. ☐ Topical mupirocin
5. ☐ Topical 1 % hydrocortisone

INCORRECT ✗

The correct answer is 2.

History and physical examination are crucial in the diagnosis of contact dermatitis. The appearance of the erythematous papules and vesicles in a linear fashion on the exposed skin suggests contact dermatitis due to plant allergens. The most common culprits are poison ivy, poison oak, and poison sumac. The chemical found in these plants, urushiol, is a very potent allergen that causes an acute dermatitis that presents as vesicles, bullae, papules, and edema. If the contact dermatitis covers more than 10 to 15% of the body surface area, oral prednisone is indicated for 14-21 days. An initial dose of 1 mg/kg/day may be used, tapered every 3 days to complete the whole course. Taking tepid baths or applying calamine lotions may promote drying when the vesicles rupture.

(Choice 1) is an antibiotic and is not indicated in this case because there is no evidence that the dermatitis is complicated with a bacterial infection.

(Choice 3) should be avoided because it may cause a secondary dermatitis that complicates the primary process. Oral diphenhydramine may be considered to treat burning and itching.

(Choice 4) is a topical antibiotic. Unless secondary bacterial infection is present, a topical antibiotic is not necessary.

(Choice 5) is a low-potency topical corticosteroid, which is ineffective in treatment of contact dermatitis. If the contact dermatitis is localized to a small area, and if it is mild, a mid-potency topical corticosteroid (e.g., mometasone furoate 0.1 % or triamcinolone acetonide 0.1 %) may be used instead of oral prednisone.

216. Question

1 points

Category: Pediatrics

A 15-year old girl presents with a 5-day history of sore throat, low-grade fever, and easy fatigability. Physical examination shows bilateral tonsillar enlargement with exudate. Her spleen is palpable 3 cm below the left costal margin. Her throat culture is negative for group A Streptococcus. Monospot test is positive. Which of the following is the most appropriate management for this patient?

- ☐ Abdominal ultrasound
- ☒ Avoidance of all contact sports
- ☐ Complete blood count
- ☐ Oral penicillin
- ☐ Splenectomy

INCORRECT ❌

The correct answer is 2.

This girl has infectious mononucleosis (IM), a common infection in adolescents caused by the Epstein-Barr virus (EBV). IM is characterized by fatigue, sore throat, fever, exudative tonsillitis, abdominal symptoms, and splenomegaly. It occurs commonly in high school and college students. Patients may have a positive Monospot test (up to 50% of patients less than 4-5 years of age may have a false negative result). EBV is usually spread in saliva and is very contagious, as suggested by the name of the disease. One of the feared complications of IM is splenic enlargement and subsequent splenic rupture, an emergency that can result in death from hemorrhagic shock. In this case, the patient has a palpable spleen 3 cm below the left costal margin, which suggests marked splenomegaly. Contact sports should be absolutely forbidden in this patient. In fact, strict bed rest, or even hospitalization, may need to be considered until the splenomegaly resolves.

(Choice 1) is usually not necessary when the diagnosis of IM is apparent and the spleen is palpable.

(Choice 3) may not help to diagnose IM. EBV infection may induce some atypical lymphocytes (which along with clinical symptoms may allow a diagnosis of IM to be made). Definitive diagnosis is made with serology.

(Choice 4) The sore throat and tonsillitis of this patient are caused by a virus (EBV), not group A Streptococcus; therefore, penicillin is not the appropriate treatment.

(Choice 5) The splenic enlargement in IM is only temporary and usually resolves on its own. Splenectomy is inappropriate at this point.

217. Question**1 points****Category: Pediatrics**

A 4-day-old infant born by spontaneous vaginal delivery without complications is brought to the pediatric emergency department. The parents report that the child is vomiting and feeding poorly. Physical examination reveals muscular rigidity with alternating bouts of flaccidity and severe opisthotonos. A bedside glucose determination shows that the infant is hypoglycemic; however, correcting the blood glucose concentration does not improve the patient's clinical condition. The urinalysis does not show evidence of infection, but the urine has a distinct odor. Other routine laboratory studies are unremarkable, except for evidence of severe acidosis. Which of the following is the most likely diagnosis?

1. ☐ Sepsis
2. ☐ Neonatal seizure
3. ☒ Maple syrup urine disease ✓
4. ☐ Trauma
5. ☐ Phenylketonuria

INCORRECT ✗

The correct answer is 3.

The abnormality in classical maple syrup urine disease (MSUD) is a deficiency of branched chain α -keto dehydrogenase, the enzyme responsible for decarboxylation the three branched-chain amino acids (B-CAAAs): leucine, isoleucine, and valine. It is characterized by the presence of high concentrations of the B-CAAAs in urine. Screening is accomplished by the addition of 2,4-dinitrophenylhydrazine (DNPH) to a sample of urine, which will cause the ketoacids of the B-CAAAs to precipitate. Additionally, the urine, skin, or hair has a characteristic smell of maple syrup or caramel. Infants present with symptoms 3–5 days after birth, these include feeding difficulties, hypoglycemia, seizures, opisthotonos, and muscular rigidity with or without intermittent flaccidity. Cortical atrophy is seen on computed tomography (CT) or magnetic resonance imaging (MRI) scans. Dietary management of intake of branched-chain amino acids is the cornerstone of therapy. However, most patients with the classical form of the disease have already suffered permanent brain damage by the time diagnosis is made and treatment is initiated. Some patients have less severe variants.

(Choice 1) A newborn with sepsis may have vomiting, poor feeding, and hypoglycemia, but the fact that this patient has a distinct odor to the urine, acidosis, no response to glucose, and bouts of flaccidity and opisthotonos makes MSUD the best answer.

(Choice 2) are a disorder of the central nervous system (CNS) that suggests hypoxic ischemic encephalopathy resulting from asphyxia, intracranial hemorrhage, hypoglycemia, infarction, or meningitis.

(Choice 4) Bruising and retinal hemorrhages might be seen if the patient had been a victim of trauma.

(Choice 5) Phenylketonuria (PKU); results from a defect in the metabolism of the amino acid phenylalanine. The infant is normal at birth, but gradually develops mental retardation. Vomiting may be an early symptom of PKU. Children with PKU are fair skinned and have eczematous rash. Some will develop a seizure disorder.

218. Question

1 points

Category: Pediatrics

A pediatrician is requested to attend the delivery of a 25-year-old primigravida, with a history of good prenatal care. The patient tells the pediatrician that she is expecting a full-term single delivery. She denies any history of sexually transmitted diseases, alcohol, or illicit drug use. Her physical examination is pertinent for a malar rash, alopecia, and polyarticular arthritis. In addition, she has a history of renal disease. This patient's newborn will have an increased risk for which of the following disorders?

1. ☒ Complete heart block ✓
2. ☐ Coarctation of the aorta
3. ☐ Open neural tube defects
4. ☐ Microcephaly
5. ☐ Polycystic kidney disease

INCORRECT ✗

The correct answer is 1.

The mother has systemic lupus erythematosus (SLE). Neonatal lupus syndrome consists of rash, thrombocytopenia, and congenital heart block. Liver disease, hemolytic anemia, and leukopenia occur less frequently. Maternal SLE is the most common cause of congenital complete heart block in the newborn. Injury is caused by maternal immunoglobulin G (IgG). The exact mechanism of damage is unknown. Most infants with congenital heart block have mothers with antibodies to Ro/SSA or La/SSB. The heart damage may be permanent. **(Choices 2,3,4 & 5)** Coarctation of the aorta, open neural tube defects, microcephaly, and polycystic kidney disease have not been associated with SLE.

219. Question

1 points

Category: Pediatrics

A term neonate is small for date and has a small head. Further physical examination of the infant demonstrates small eyes with short palpebral fissures, a flattened nose, and abnormal palmar creases. With which of the following maternal conditions is this presentation most likely associated?

1. ☒ Alcohol abuse ✓
2. ☐ Cirrhosis
3. ☐ Cocaine abuse
4. ☐ Diabetes mellitus
5. ☐ Hypothyroidism

INCORRECT ✗

The correct answer is 1.

Maternal alcohol abuse is a common cause of teratogenesis. The fetal alcohol syndrome is characterized by potentially severe mental retardation secondary to impaired brain development. Other phenotypic features seen in severely affected infants include growth retardation, microcephaly, microphthalmia, short palpebral fissures, and midfacial hypoplasia. In addition, these infants may have abnormal palmar creases, cardiac defects, and joint contractures. Partial expression in mild cases is common and may make diagnosis difficult.

(Choice 2) Maternal cirrhosis can decrease fertility and cause miscarriages and prematurity, but does not specifically cause fetal malformation.

(Choice 3) Maternal cocaine abuse can cause spontaneous abortion, fetal death, placental abruption, low birth weight, reduced body length, and reduced head circumference.

(Choice 4) Maternal diabetes mellitus can cause large for-gestational-age infants who tend to be hypoglycemic after birth.

(Choice 5) Maternal hypothyroidism may worsen during pregnancy but usually does not produce prominent fetal effects.

220: Question

1 points

Category: Pediatrics

A 6-year-old girl who appears healthy is brought to the primary care physician by her mother because of a rash. The mother states that the patient had been in her usual state of good health until 2 days earlier, when she developed a tactile fever and mild upper respiratory symptoms. Yesterday, the patient had erythematous facial flushing that spread as a macular red lesion to her proximal extremities and trunk, which now has a lacy appearance. Which of the following is the most likely diagnosis?

1. ☒ Erythema infectiosum ✓
2. ☐ Roseola
3. ☐ German measles
4. ☐ Measles
5. ☐ Scarlet fever

INCORRECT ✗

The correct answer is 1.

Erythema infectiosum (also known as Fifth disease) is a mild exanthematous disease caused by parvovirus B19. Usually, there is no prodrome, and fever is low-grade or absent. The rash usually starts on the cheeks, which appear to have been slapped. Following the bright-red and confluent rash, an erythematous maculopapular rash appears on the trunk, although it can precede the facial rash. The rash then fades, giving a lacy or reticular appearance. The rash is often pruritic, but does not desquamate, as seen with measles. Other symptoms include headache, rhinitis, and arthralgia, but these symptoms are more common in adults.

(Choice 2) In most cases, roseola is caused by herpesvirus 6. Children with roseola present with the differential diagnosis of a fever of unknown origin until the temperature drops precipitously and the rash appears. The rash is maculopapular and nonpruritic, and it blanches on pressure. It first appears on the trunk and spreads to the arms and neck (mild on legs and face).

(Choice 3) Rubella (also known as German measles, or three-day measles, is another viral exanthematous disease of children and adults, with worldwide distribution. The signs and symptoms include adenopathy (posterior auricular), low-grade fever, arthralgias, coryza, and conjunctivitis. The exanthem is descending, beginning on the face or neck and spreading in hours to the trunk and extremities. The lesions are maculopapular and may desquamate. The rash fades in 24–48 hours.

(Choice 4) is a highly contagious viral infection transmitted by respiratory droplets. Prodromal symptoms include brassy cough, coryza, conjunctivitis, and fever. Koplik's spots (grayish white spots with a red halo) appear on the buccal mucous membrane 24–48 hours before the rash. The rash begins on the face and ears and spreads to the trunk and extremities in 24–36 hours. The lesion is maculopapular and may become confluent on the face and body (a distinct characteristic of measles).

(Choice 5) is an upper respiratory infection associated with a characteristic diffuse maculopapular "sandpaper"-type rash and group A streptococcal pharyngitis. The face is usually spared, but the cheeks may be erythematous and the area around the mouth spared. Pastia's lines (prominence of the rash in the creases of the elbow, axillae, and groin) and strawberry tongue (in which the tongue is coated and the papillae may be swollen and reddened) are also associated with scarlet fever.

221. Question

1 points

Category: Pediatrics

A 10-year-old girl is involved in a motor vehicle accident, sustaining multiple injuries to her head, arms, and abdomen. Her blood pressure is 90/160 mm Hg, and her pulse is 120/min. Her forearm is disfigured, and bone can be seen through the wound. She is breathing periodically and has cyanotic lips. Her abdomen is rigid, and there is flank discoloration. Which of the following is the most appropriate next step in management?

- ☐ Splint the arm and cover wound with sterile gauze
- ☒ Administer crystalloid solution ✓
- ☐ Administer vasopressors immediately
- ☐ Administer packed red blood cells
- ☐ Perform exploratory laparotomy

INCORRECT ✗

The correct answer is 2.

The patient is losing blood, most likely in the retroperitoneum, as a result of traumatic injury. Her blood pressure is decreased, and she is compensating with an increased pulse. The immediate response is repletion of fluids to counter the hypovolemic shock the patient is experiencing. Perfusion to the brain and heart is of utmost importance.

(Choice 1) The broken arm will need to be corrected after she has been surgically stabilized. The emergency option is to splint the arm. However, an orthopedic surgeon should evaluate for nerve and vessel damage as well.

(Choice 3) Although administration of vasopressors may correct the blood pressure, it is not addressing the underlying problem. The patient has lost blood volume, and pressors will simply clamp down blood supply to the viscera and limbs, leading to ischemic damage. Fluid should be given first, with pressors as an adjunct.

(Choice 4) The patient is probably losing large quantities of blood and will need replenishment soon. However, the blood should not be replenished too quickly, as this may lead to worsening shock. Thus, the patient should first be stabilized with fluids; then, blood should be replenished.

(Choice 5) The underlying problem is a retroperitoneal bleed, and exploratory laparotomy will be needed to correct the problem. This is a surgical treatment, however, and it is ideal to stabilize the patient's hemodynamics before the surgical correction.

222. Question

1 points

Category: Pediatrics

A father brings his 18-month-old daughter to the office for the evaluation of fever for the past 10 days. The father, who works as a phlebotomist, says that the temperature seems higher in the morning and again at night, often reaching 40.5 °C (105 °F), and is sometimes associated with shaking chills. He remembers that the child developed severe respiratory distress and skin rash when treated with penicillin in the past. He has heard about the development of 'liver toxicities' in children treated with aspirin, and he is interested in knowing everything about his daughter's condition. On examination, the child is in no acute distress. Her temperature is 39.4 °C (103 °F), pulse rate is 90/min, and blood pressure is 100/60 mm Hg. Her sclerae are anicteric. There is bulbar conjunctival injection, and the oropharynx shows diffuse mucosal injection with fissuring and crusting of the lips. There are numerous hyperemic macules over her entire body, and her palms appear edematous. Based on the above description, which of the following is the most accurate statement?

1. ☐ Latex agglutination test is more accurate than streptococcal throat test in determining the etiology of this condition
2. ☐ Immediate desensitization & treatment with penicillin V will prevent further renal complications
3. ☐ Thrombocytopenia is an important marker in the second week of this illness
4. ☐ Acetaminophen is the drug of choice for alleviation of high temperatures in this child
5. ☒ She needs to be hospitalized and treated with intravenous immune globulin (IVIG) and aspirin ✓

INCORRECT ✗

The correct answer is 5.

The case in question satisfies the criteria of Kawasaki's disease, which are:

1. Fever for > 5 days.
2. Four of the following symptoms:
 - Bulbar conjunctival injection
 - Desquamation of the finger and toe tips, indurative edema
 - Erythema, fissuring, and crusting of the lips, strawberry tongue, and diffuse mucosal injection of the oropharynx
 - Morbilliform truncal exanthem
 - Cervical lymphadenopathy

Kawasaki disease or mucocutaneous lymph node syndrome is one of the most common causes of generalized vasculitis in children. It is usually self-limited, although it can be fatal because of giant aneurysm formation, thrombosis or rupture of the coronary arteries, leading to myocardial infarction. 2D echocardiography is used to assess cardiac function and coronary vasculature. A baseline echocardiography is to be performed within 7 days of the disease onset, and then repeated 6 to 8 weeks later.

(Choice 1) The most common condition with which Kawasaki's disease is confused is scarlet fever; however, scarlet fever will have a positive streptococcal throat test and normal-appearing lips, as opposed to negative streptococcal throat test and inflamed lips in Kawasaki's disease. Latex agglutination test is more rapid but less accurate in the diagnosis of scarlet fever.

(Choice 2) The treatment of choice for scarlet fever is a ten-day course of penicillin V; however, in penicillin-allergic patients, erythromycin or clindamycin can be used. Treatment of scarlet fever with penicillin doesn't prevent post-streptococcal glomerulonephritis.

(Choice 3) The laboratory abnormalities associated with Kawasaki's disease are increased acute phase reactants such as erythrocyte sedimentation rate (ESR) and C-reactive protein, leukocytosis, normochromic-normocytic anemia, and thrombocytopenia. Thrombocytopenia is a non-specific finding and does not characterize this illness.

(Choice 4) Fever in Kawasaki's disease poorly responds to acetaminophen. All patients with suspected Kawasaki disease should be hospitalized and treated with intravenous immune globulin (IVIG) and high-dose aspirin. The mainstays of systemic treatment of Kawasaki's disease are intravenous immunoglobulin (IVIG) and aspirin. Aspirin is useful for fever and arthritis. IVIG has been shown to reduce the incidence of coronary artery aneurysms and other complications. If untreated, up to 25 percent of these patients may develop coronary artery aneurysms.

223. Question

1 points

Category: Pediatrics

An 8-year-old girl is brought to the pediatrician's office for evaluation of new onset swelling around the eyes. Physical examination reveals periorbital, sacral, and pretibial edema; her blood pressure is 96/64mm Hg. The rest of her physical examination is normal. Which of the following is the most appropriate initial diagnostic study?

1. ☐ Levels of liver enzymes
2. ☐ Radiography of the chest
3. ☐ Transthoracic echocardiography
4. ☐ Ultrasonography of the kidneys
5. ☒ Urinalysis ✓

INCORRECT ✗

The correct answer is 5.

Apparently, the girl in this clinical vignette has generalized edema (i.e., anasarca). The most common causes of swelling are nephrotic syndrome, congestive heart failure, and liver disease. Other causes include hypothyroidism, shock, mineralocorticoid excess, and malnutrition. Since there is no evidence of any abnormal cardiac or hepatic findings, urinalysis is the most appropriate initial diagnostic test to screen for proteinuria. In children with nephrotic syndrome, albumin (a serum protein) is lost in the urine and hence causes the oncotic pressure inside the vessels to decrease. Fluid then shifts from the intravascular space into the interstitial space, causing generalized edema.

(Choices 1,2,3 & 4) Liver enzymes, chest radiographs, echocardiography, and kidney ultrasound are not indicated in this case as the initial diagnostic test.

224. Question

1 points

Category: Pediatrics

A 7-year-old girl is brought to the physician because of an exanthematous rash associated with malaise and headache for 2 days. On examination, the child shows a fiery red facial rash with a characteristic "slapped cheek" pattern and pallor around the mouth. There is no fever. In immunocompromised patients, the pathogen that causes this condition may result in which of the following manifestations?

1. ☒ Aplastic anemia ✓
2. ☐ Encephalitis
3. ☐ Non-Hodgkin lymphoma
4. ☐ Progressive multifocal leukoencephalopathy (PML)
5. ☐ Symmetric polyarthritis

INCORRECT ✗

The correct answer is 1.

Parvovirus B19 is the etiologic agent of this benign exanthem of childhood, which manifests with a "slapped cheeks" facial rash and little or no fever. This exanthema is referred to as fifth disease or erythema infectiosum. B19 parvovirus may cause red cell aplasia (aplastic anemia) in patients with AIDS or with sickle cell disease. Older people develop symmetric polyarthritis (**Choice 5**), which is not seen in children.

(**Choice 2**) in immunocompromised patients may have different etiologies. Herpes simplex 1 and 2, herpes zoster, measles, and cytomegalovirus may cause encephalitis in immunocompromised patients. HIV itself frequently affects the CNS, producing subacute encephalitis in AIDS patients.

(**Choice 3**) in immunocompromised patients, especially those with AIDS, is most commonly a high-grade B-cell lymphoma Epstein-Barr virus has been implicated in its pathogenesis.

(**Choice 4**) is a complication of JC virus, a papovavirus that causes no disease in immunocompetent individuals. PML affects patients with AIDS or lymphomas. It is a progressive demyelinating disease in which oligodendroglial cells are destroyed by the virus.

225. Question

1 points

Category: Pediatrics

A 5-year-old boy is brought to clinic with increasing right lower foot pain. He stepped on a nail several days ago. At that time, the family had sought medical attention. The child was given a tetanus shot, and the wound was extensively irrigated. On examination, the foot is tender, swollen, warm, and erythematous. Osteomyelitis is suspected. Which of the following is the most appropriate next step in diagnosis?

1. ☐ White cell count
2. ☐ CT scan of the foot
3. ☐ Gallium scan
4. ☒ Technetium bone scan
5. ☐ X-ray of the foot

INCORRECT ❌

The correct answer is 4.

A technetium bone scan is very effective in diagnosing osteomyelitis in its earliest stages. Technetium uptake reflects osteoblastic activity and skeletal vascularity, so this method is quite sensitive. However, it will give false positive results for fractures, tumors, infarction, or neuropathic osteopathy, and will give false negative results if the vascular supply is impeded in some way.

(Choice 1) is helpful in monitoring the progress and response to treatment but does not aid in the absolute diagnosis of osteomyelitis.

(Choice 2) would not be as sensitive as an MRI scan, and a technetium scan would be better than either.

(Choice 3) are effective in demonstrating response to treatment, but not in the initial diagnosis of osteomyelitis.

(Choice 5) will not show osteomyelitis for approximately 1 week.

226. Question

1 points

Category: Pediatrics

A female infant develops cyanosis. She was delivered vaginally at 43 weeks' gestation, and the amniotic fluid had a greenish color. She has dry, peeling skin, emaciated extremities and long fingernails. Tachypnea, nasal flaring, and sub- and intercostal retractions are observed. Auscultation reveals rales and rhonchi. The chest radiograph reveals flattening of the diaphragm, hyperinflation of the lungs, and patchy opacities diffusely distributed over both lung fields. Arterial blood gases reveal hypoxia and hypercarbia. What is most likely impairing the normal respiratory function of this newborn?

1. ☐ Fetal lung fluid
2. ☐ Poor alveolar compliance
3. ☒ Meconium ✓
4. ☐ Paralysis of the diaphragm

INCORRECT ✗

The correct answer is 3.

The patient's history and physical findings are suggestive of meconium aspiration. The greenish amniotic fluid indicates the presence of meconium. The condition is more common in post-mature and small-for-gestational-age (SGA) infants. The characteristic presentation includes signs of respiratory distress (i.e., marked tachypnea and cyanosis at birth), rales, rhonchi, nasal flaring, grunting, and use of accessory muscles of respiration (e.g., subcostal and/or intercostal retractions). Hyperinflation occurs secondarily to the valve-like mechanism by which meconium obstructs the airways, keeping air from exiting the lungs after each inspiration. This state of hyperinflation consequently leads to a barrel-shaped chest (increased antero-posterior diameter). Hypoxia and acidosis also ensue, thus increasing pulmonary vascular resistance and aggravating the respiratory compromise. The typical chest radiograph of meconium aspiration (as seen in this patient) includes flattening of the diaphragm, hyperinflation, coarse streaking and patchy opacities over both lung fields. Complications include pneumothorax, infection, pulmonary hypertension, residual lung problems and permanent neurological impairment due to asphyxia.

(Choice 1) Fetal lung fluid causes respiratory difficulty in transient tachypnea of the newborn. This condition is characterized by lack of clearance of fetal lung fluid and occurs most commonly in fetuses delivered by cesarean section. It is a self-limited condition that generally improves after a few hours. Auscultation does not show rales or rhonchi. The lungs are usually very clear. Patients usually do not require more than 40% FIO₂ to oxygenate adequately.

(Choice 2) Poor alveolar compliance is characteristic of respiratory distress syndrome. It is secondary to lack of pulmonary surfactant and occurs primarily in preterm infants. This patient is post-mature, based on the gestational age, dry and peeling skin, emaciated extremities and long fingernails.

(Choice 4) The flattening of this patient's diaphragm is due to hyperinflation (not paralysis) of the lungs. Furthermore, the characteristic radiographic finding of diaphragmatic paralysis is an elevated hemi-diaphragm.

227. Question

1 points

Category: Pediatrics

A premature neonate with respiratory distress syndrome is maintained on mechanical ventilation in a neonatal intensive care unit. Two weeks after delivery, the nurses in the intensive care unit notice that higher ventilation settings are needed and that more secretions are being suctioned from the endotracheal tube. A chest x-ray film shows questionable new infiltrates. Which of the following is the most likely pathogen?

1. ☒ Coagulase-negative oxacillin-resistant *Staphylococcus* ✓
2. ☐ Coagulase-negative oxacillin-sensitive *Staphylococcus*
3. ☐ Group B *Streptococcus*
4. ☐ Methicillin-resistant *Staphylococcus aureus*
5. ☐ Methicillin-sensitive *Staphylococcus aureus*

INCORRECT ✗

The correct answer is 1.

This infant has "late-onset" infant pneumonia, since it occurred more than 1 week after delivery. The most common cause in this setting is coagulase-negative *Staphylococcus*, which in this setting is usually resistant to oxacillin. Cultures of blood and tracheal aspirates can usually demonstrate the organism. Vancomycin is the initial drug of choice and may sometimes be replaced by a less toxic drug after culture sensitivities become available.

(Choice 2) Most strains of coagulase-negative *Staphylococcus* isolated in this setting are resistant, rather than sensitive, to oxacillin.

(Choice 3) is an important cause of early onset neonatal sepsis and pneumonia.

(Choices 4 & 5) Methicillin-sensitive and methicillin-resistant *Staphylococcus aureus* tend to cause skin pustules of the periumbilical and diaper areas.

228. Question

1 points

Category: Pediatrics

A 4-month-old infant is brought to the emergency department with sudden onset of lethargy and poor feeding. The mother reports that the infant had been in her usual state of good health until recently, when she developed constipation. The mother also states that she has recently stopped breastfeeding and has changed to formula feeding. According to the mother, the infant does not like the formula unless it is sweetened; hence, she adds a teaspoonful of honey. On physical examination, the infant is noted to be afebrile and have generalized hypotonia and weakness, as well as ophthalmoplegia. Which of the following is the most likely diagnosis?

1. ☐ Guillain-Barré syndrome
2. ☐ Hypothyroidism
3. ☐ Werdnig-Hoffmann disease
4. ☒ Infant botulism ✓
5. ☐ Hypomagnesemia

INCORRECT ✖

The correct answer is 4.

Infant botulism usually occurs in infants younger than 1 year of age, particularly between 2 and

6 months. It occurs after the ingestion of *Clostridium botulinum* spores, which can be found in honey and corn syrup, as well as in soil and house dust. The course of the disease varies but progresses quickly. Typically, the child is afebrile and has been previously healthy. The infant becomes constipated and feeds poorly. A weak cry, hypotonia, and loss of head control are then seen. Descending paralysis progresses over hours to days. Ventilatory support may be necessary.

(Choice 1) consists of weakness that begins in the lower extremities and spreads to the trunk, the upper extremities, and the bulbar muscles. This pattern is known as Landry ascending paralysis, and respiratory failure may result.

(Choices 2 & 3) Although hypothyroidism and Werdnig-Hoffmann disease present with hypotonia, it is seen earlier in life, and progression of symptoms occurs more slowly.

(Choice 5) usually presents in conjunction with hypocalcemia and manifests as tetany.

229. Question

1 points

Category: Pediatrics

An 8-year-old boy is brought to the pediatrician with a rash on his abdomen. The mother first noticed the rash about 3 weeks ago. The boy has no fever or other symptoms. On examination, there is a well-circumscribed, circular, erythematous, scaly annular patch on his abdomen. The border of the skin lesion is raised and well defined. Which of the following is the most likely diagnosis?

1. ☐ Erythema multiforme
2. ☐ Erythema nodosum
3. ☐ Impetigo
4. ☐ Nummular eczema
5. ☒ Tinea corporis ✓

INCORRECT ✗

The correct answer is 5.

The description of this skin lesion is consistent with tinea corporis, a very common cutaneous infection in children. It is one of the dermatophytoses (ringworm), which is caused by a group of related fungi including *Microsporum* and *Trichophyton* species. These fungi can invade the stratum corneum, nails, and hairs. The lesions are annular, gyrate, scaling, and discrete. Secondary bacterial infection commonly occurs. Topical antifungal agents are the initial treatment of choice. Tinea pedis represents the same infection in the lower extremities, whereas tinea manuum affects the hands.

(Choice 1) is a skin eruption caused by a hypersensitivity reaction to drugs, infections, or toxins. Skin lesions usually appear as macules, papules, wheals, and vesicobullous lesions. There is often central clearing of those skin lesions, which are therefore termed "target lesions."

(Choice 2) is characterized by tender erythematous nodules, usually in the pretibial surfaces. It is caused by hypersensitivity reactions to a spectrum of infections, drugs, and disease states.

(Choice 3) is a superficial bacterial infection of the skin usually caused by group A streptococci. Infection is usually induced by minor trauma or abrasion to the surface of the skin with prior colonization of the bacteria.

(Choice 4) is a highly pruritic idiopathic skin disorder that commonly occurs in association with asthma or allergic rhinitis.

230. Question

1 points

Category: Pediatrics

A newborn infant is brought to the nursery for evaluation after delivery. The mother reports that the pregnancy was uncomplicated, but she had only two prenatal visits. The infant was born via normal spontaneous vaginal delivery and required no resuscitation. The infant has hepatosplenomegaly on examination. While in the hospital, the infant requires treatment for anemia and hyperbilirubinemia. On subsequent examinations, the infant has clear rhinorrhea and ulcerative lesions on his feet. Which of the following congenital infections is most likely in this patient?

1. ☐ Toxoplasmosis
2. ☒ Syphilis
3. ☐ Rubella
4. ☐ Cytomegalovirus infection
5. ☐ Human immunodeficiency virus infection

INCORRECT ❌

The correct answer is 2.

Congenital syphilis is contracted through the transplacental transmission of *Treponema pallidum*. Most infants born with congenital syphilis are initially asymptomatic. Those with early symptoms present with cutaneous lesions on the palms and soles, hepatosplenomegaly, jaundice, anemia, and rhinorrhea. Radiographs demonstrate metaphyseal dystrophy and periostitis. Late congenital manifestations (Presenting after two years of age) include frontal bossing, high arched palate, Hutchinson teeth, interstitial keratitis, saddle nose, and perioral fissures. These late manifestations can be prevented with early treatment of the infant.

Diagnosis of syphilis is made with serologic testing. The initial screening is with nontreponemal tests such as the venereal disease research laboratory (VDRL) test, rapid plasma reagin (RPR) test, or enzyme immunoassay (EIA) test; confirmatory testing is with treponemal tests such as the fluorescent treponemal antibody absorption (FTA-ABS) test or the treponema pallidum particle agglutination assay (TPPA). Parenteral penicillin G is the treatment of choice for syphilis.

(Choice 1) Major clinical findings of congenital toxoplasmosis include hepatosplenomegaly, hydrocephalus, chorioretinitis, and intracranial calcifications.

(Choice 3) Congenital rubella presents with sensorineural hearing loss, cataracts, heart defects, hepatosplenomegaly, microcephaly, and thrombocytopenic purpura (commonly described as a "blueberry muffin" rash).

(Choice 4) Congenital cytomegalovirus is characterized by intrauterine growth retardation, hepatosplenomegaly, petechiae or purpura, microcephaly, chorioretinitis, sensorineural hearing loss, and periventricular calcifications.

(Choice 5) Infants with congenital human immunodeficiency virus (HIV) infection are usually clinically asymptomatic at birth.

231. Question

1 points

Category: Pediatrics

The mother of a 2-year-old boy comes to the physician because her child awakens at night, with a blank gaze, screaming in bed without recognizing his parents. These episodes have occurred three times in the past 2 weeks, always in the first few hours of the night. The child goes back to sleep and seems to retain no memory of the episode the next morning. Which of the following is the most appropriate next step in management?

1. ☒ Reassurance of parents about the nature of these manifestations ✓
2. ☐ Avoidance of TV before going to bed
3. ☐ Behavioral therapy
4. ☐ Therapy with chloral hydrate
5. ☐ Therapy with a tricyclic antidepressant

INCORRECT ✗

The correct answer is 1.

The most common disorders of sleep in childhood include difficulty falling and staying asleep, night terrors, nightmares, and sleepwalking. The clinical presentation in this case is consistent with night terrors. These usually begin after 18 months of age and affect as many as 6% of children. The frequency of episodes is highly variable. Night terrors occur in the first few hours after falling asleep in concurrence with the transition from non-REM to REM sleep. Night terrors will usually disappear in late childhood. There is evidence that this is a genetic disorder, since it is frequently associated with a family history of sleep disturbances, including night terrors, sleepwalking, and sleep-talking. Management of this condition should simply be based on explaining the benign nature of this disorder to the parents. No other measure is necessary besides preventing physical injury during the episodes if the child is very agitated.

(Choice 2) has no benefit in preventing night terrors. Nightmares, on the other hand, may be triggered by frightening TV shows. Nightmares occur in the last part of sleep, during the REM phase. The child appears frightened during nightmares, but recognizes his caretakers and is easily consoled. After nightmares, children may not fall back asleep easily, as happens after night terrors.

(Choices 3 & 4) Behavioral therapy and pharmacologic therapy with chloral hydrate have no value in either night terrors or nightmares. Short-term treatment (1-2 weeks) with chloral hydrate may be tried in children with problems falling asleep.

(Choice 5) Benzodiazepines or tricyclic antidepressants have been used occasionally for night terrors and nightmares, based on their suppression of stage 3 and 4 sleep, but studies confirming the efficacy of these agents is lacking.

232. Question

1 points

Category: Pediatrics

A 16-month-old girl is brought to medical attention because of irritability, poor feeding, and temperatures up to 39.4 C (103 F). Careful history and physical examination fail to disclose any identifiable cause of her fever. There is some degree of abdominal tenderness on palpation. Which of the following is the most appropriate next step in diagnosis?

1. ☐ Microscopic examination and culture of stool
2. ☐ Renal ultrasound
3. ☐ Chest x-ray films
4. ☐ Voiding cystourethrogram
5. ☒ Culture of urine obtained by transurethral catheterization ✓

INCORRECT ✗

The correct answer is 5.

Urinary tract infections (UTIs) are extremely common in children, especially girls. Failure to diagnose and properly treat UTIs may lead to serious complications, including renal scarring, hypertension, and chronic renal failure. Renal scarring due to UTIs is one of the most common causes of renal failure in children and young adults. UTIs in children younger than 2 years usually present with nonspecific signs and symptoms. Irritability, poor feeding, diarrhea, and fever are the most common manifestations. Occasionally, abdominal tenderness and malodorous urine are present. A diagnosis of UTI becomes more likely in a child with fever (especially if the temperature is higher than 39 C) but no identifiable sources of infection. In such cases, urinalysis and urine culture are absolutely necessary to diagnose UTI. Bagged urine specimens are frequently associated with false positive results because of bacterial contamination. Transurethral catheterization is the most widely recommended method of urine collection for UTI screening. Urinalysis and culture are the most important diagnostic investigations in children younger than 2 years who present with unexplained fever.

(Choices 1 & 3) Microscopic examination and culture of stool or chest x-ray films should not be the first diagnostic investigations in cases of fever without an obvious source. Of course, careful history and physical examination are crucial to rule out causes of fever other than UTIs.

(Choice 2) is now considered the method of choice to identify obstructive lesions that may predispose to UTIs. Renal ultrasound is performed after a diagnosis of UTI is established.

(Choice 4) is the most sensitive technique to identify vesicoureteral reflux in children who have had an episode of pyelonephritis.

233. Question

1 points

Category: Pediatrics

A 10-year-old boy is brought to clinic because of increasing weakness and dyspnea over the past 6 months. He has been previously healthy and is on no medications. There is no significant family history of illness. On examination, he appears pale. His hematocrit is 20%, mean corpuscular volume (MCV) is $60/\mu\text{m}^3$, and iron level is normal. Smear reveals basophilic stippling. Hemoglobin electrophoresis is consistent with the absence of one β -globin gene. Which of the following is the most likely diagnosis?

1. ☐ Elliptocytosis
2. ☐ Hemoglobin S-C disease
3. ☐ Porphyria
4. ☐ Sickle cell disease
5. ☒ Thalassemia 

INCORRECT 

The correct answer is 5.

Thalassemia refers to any genetic defect in the production of the globin chains of hemoglobin. Patients may have deficient production of the globin β -chain (β -thalassemia) or α -chain (α -thalassemia). This patient has the heterozygous β -thalassemia minor condition. In this condition, electrophoresis would show an elevated percentage of hemoglobin A1 or hemoglobin F. This type of thalassemia differs from iron deficiency anemia in that basophilic stippling is present in red cells caused by retention of ribonucleoprotein particles and mitochondrial remnants in the red cells.

 **(Choice 1)** is similar to spherocytosis in that it is caused by a defect in a cell wall structural protein and will lead to elliptical red cells on the peripheral blood smear.

(Choice 2) is a variant of sickle disease in which the cells carry two abnormal β -alleles, those for hemoglobin S and those for hemoglobin C. Such patients are sometimes severely affected, but generally less so than those with HbSS disease.

(Choice 3) is due to a defect in heme biosynthesis. Each of the heme biosynthetic enzymes is encoded by a separate gene on chromosomes 1, 3, 9, 14, and 18, among others.

(Choice 4) is due to a structural defect in hemoglobin that leads to sickling of red cells in conditions of low oxygen tension, low flow, and decreased blood vessel diameter, leading to hemolysis.

234. Question

1 points

Category: Pediatrics

A 5-year-old girl is diagnosed with Bordetella pertussis infection after an outbreak in the day-care center. She is prescribed erythromycin for 14 days. She lives with her parents and 13-year-old sister whose immunizations are up to date. What is the most appropriate way to limit the risk of infection in household contacts?

1. ☐ Administer pertussis immunization to all household contacts
2. ☒ Prescribe erythromycin for 14 days to all household contacts
3. ☐ Prescribe erythromycin for 14 day to the mother and father only
4. ☐ Administer pertussis immunization to the parents only
5. ☐ Keep the child hospitalized until symptoms disappear

INCORRECT ❌

The correct answer is 2.

Pertussis is a highly contagious disease despite immunization. Transmission occurs via respiratory droplets. Infection and vaccination both confer only transient protection. However, despite immunization, over 75% of household contacts will develop some symptoms of pertussis if exposed. The recommended first line treatment for pertussis is a macrolide antibiotic such as erythromycin, azithromycin, or clarithromycin. Treatment during the catarrhal stage may help shorten the disease duration. Treatment after the catarrhal stage will not shorten the disease course, but will help decrease transmission. Due to the high rate of transmission, chemoprophylaxis is recommended for all close contacts despite their vaccination status. Macrolide antibiotics are the preferred prophylactic treatment. In addition to prophylaxis, close contacts should receive the pertussis vaccination according to the recommended immunization schedule if they are not already fully immunized.

(Choices 1 & 4) Pertussis immunization now involves an acellular vaccine, since this has been associated with less adverse effects than whole-cell vaccine. It is given to all children in 5 doses along with diphtheria and tetanus vaccines. Three doses are given in infancy (at 2, 4 and 6 months), one dose between 15 and 18 months, and the last one between 4 and 6 years. In children <7 years of age who have received less than 4 doses of pertussis vaccines, an additional dose is given to complete the recommended series.

(Choice 5) The treatment of pertussis is erythromycin for 14 days. Respiratory isolation is maintained only during the first 5 days of erythromycin therapy. Hospitalization is indicated in infants < 3 months, those between 3 to 6 months with severe paroxysms, and if significant complications occur. Although albuterol and corticosteroids have been used to reduce the severity of symptoms, no rigorous trials have been conducted to confirm their effectiveness.

235. Question

1 points

Category: Pediatrics

A 2-year-old boy presents to the emergency department (ED) with severe wheezing. His symptoms have not improved even after three treatments with nebulizers. This is his third trip to the ED in the past two weeks. In the ED, he is given a dose of intravenous steroids, but continues to wheeze. On auscultation, there is no air entry bilaterally. His chest x-ray result is normal. His oxygen saturation is 91 % on 6 liters of oxygen. What is the next best step in the management of this patient?

1. ☐ Start theophylline
2. ☐ Tracheostomy
3. ☒ Mechanical ventilation 
4. ☐ Racemic epinephrine
5. ☐ Intravenous antibiotics

INCORRECT 

The correct answer is 3.

Asthma is characterized by airway hyperreactivity, bronchospasm and inflammation. It is a common illness in childhood, and approximately 10% of children present to the ED with unremitting asthma (i.e., continuous wheezing despite adequate treatment). Of these patients, fewer than 10% require mechanical ventilation; however, hospitalization is mandatory. In this case, the child has a "silent chest" or absent air entry, and continues to desaturate despite prednisone treatment; therefore, the best option is hospitalization and mechanical ventilation.

(Choice 1) Theophylline is an option when wheezing is severe; however, the agent must be given intravenously and levels must be monitored since the drug has a tendency to increase the heart rate. For acute status asthmaticus, theophylline does not help much.

(Choice 2) Tracheostomy is a surgical procedure when the upper airways are obstructed. It is nearly always an elective procedure, and never done to relieve asthma.

(Choice 4) Racemic epinephrine may help a little in moderate asthma, but when the condition is severe, its administration will just delay the initiation of mechanical ventilation. Racemic epinephrine has a better role in the management of croup.

(Choice 5) Antibiotics are not indicated if there is no evidence of infection.

236. Question

1 points

Category: Pediatrics

A 7-year-old boy is brought to your office with a mild fever and neck swelling. His mother says that the boy has been complaining of neck pain for a couple of days and she noticed a tender neck lump yesterday. All of his vaccinations are up-to-date. He has no known allergies. Physical examination reveals a tender and fluctuant anterior cervical mass. Which of the following is the best medication for this patient?

1. ☐ Penicillin
2. ☐ Amoxicillin
3. ☒ Dicloxacillin ✓
4. ☐ Erythromycin
5. ☐ Acyclovir

INCORRECT ✗

The correct answer is 3.

Rapidly enlarging, fluctuant cervical lymph nodes typically represent streptococcal or staphylococcal infections, especially in children. The treatment is incision and drainage of the mass plus an antibiotic active against these two types of organisms. Of the antibiotics listed, dicloxacillin covers streptococci and has the best anti-staphylococcal coverage. Of note, dicloxacillin will treat infections caused by methicillin-sensitive *Staphylococcus aureus* (MSSA) but not methicillin-resistant *Staphylococcus aureus* (MRSA).

(Choice 1) Penicillin is a β -lactam antibiotic effective in treating *Listeria* and many streptococcal infections. However, staphylococci produce the β -lactamase enzyme, which renders penicillin and aminopenicillins ineffective. Therefore, penicillin is a poor choice when there is a high likelihood of *S. aureus* infection. 

(Choice 2) Like penicillin, amoxicillin is susceptible to inactivation by β -lactamase and should not be used when *S. aureus* infection is likely.

(Choice 4) Erythromycin is a macrolide antibiotic that is among the first-line agents for treating streptococcal pharyngitis and mild community acquired pneumonia. It may have activity against *S. aureus*, but is not a reliable anti-staphylococcal drug.

(Choice 5) Acyclovir is an antiviral medication used to treat infections caused by viruses in the herpes family. It has no role in treating patients with streptococcal or staphylococcal infections.

237. Question

1 points

Category: Pediatrics

A 7-year-old boy has a history of repeated urinary tract infections that have been treated by the empiric use of antibiotics. The parents are not satisfied with the care the child is receiving, and they take him to a pediatric urologist. Evaluation by voiding cystourethrogram shows that the patient has vesicoureteral reflux without ureteral or upper tract dilatation (grade one reflux). Which of the following is the appropriate management for this child?

1. ☐ Alpha-blockers
2. ☒ Long-term, low-dose antibacterial therapy ✓
3. ☐ Nephrectomy on the affected site
4. ☐ Reassurance and observation
5. ☐ Surgical reimplantation of the ureter

INCORRECT ✗

The correct answer is 2.

Low-grade reflux can be expected to resolve as the child grows, and the ureteral implantation into the bladder becomes more oblique. While waiting for that to happen, however, the kidney must be protected against infection. Long-term, low dose antibacterial therapy is thus recommended.

(Choice 1) The problem is caused by an abnormally short intramural ureteral tunnel rather than bladder neck obstruction. The use of α -blockers is thus not indicated.

(Choice 3) The goal of therapy is to protect and preserve renal function. To do a nephrectomy would do the opposite.

(Choice 4) Spontaneous resolution is expected, but simple reassurance and observation would leave the kidney unprotected.

(Choice 5) Surgical correction is needed in more severe cases in which spontaneous resolution is not likely to occur.

238. Question

1 points

Category: Pediatrics

A 3-year-old-boy ingests 40 of his older sister's chewable vitamin tablets, as well as 3 tablets of 250 mg of acetaminophen. The ingredients in the multivitamin tablets are as follows:

Vitamin A: 3000 IU

Thiamine: 1 mg

Vitamin C: 75mg

Vitamin B₆: 1 mg

Vitamin D: 400IU

Iron: 12mg

Fluoride: 1 mg

The child is brought to the emergency department in no acute distress. Which of the following complications may occur if appropriate therapy is not undertaken?

1. ☐ Acute renal failure from vitamin D toxicity
2. ☐ Hepatic failure from acetaminophen toxicity
3. ☒ Hepatic failure from iron toxicity ✓
4. ☐ Increased intracranial pressure from vitamin A toxicity
5. ☐ Intestinal ischemia from fluoride toxicity

INCORRECT ✖

The correct answer is 3.



Vitamin A, iron, and fluoride can cause acute toxic reactions. Iron is an important ingredient in chewable vitamins. Toxic effects occur after ingestion of 30 mg/kg of body weight and include acute corrosive necrosis of the stomach and acute hepatic necrosis. Deferoxamine, an iron chelator, may be used in treatment.

(Choice 1) Vitamin D has no significant acute toxicity in the amounts available in chewable tablets, but chronic overuse may cause renal failure.

(Choice 2) The most serious toxicity of acetaminophen toxicity is irreversible hepatotoxicity, which may lead to death. If the presumed ingested dose is less than 100 mg/kg of body weight, the ingestion is mild and need not be treated.

(Choice 4) Hypervitaminosis A occurs in amounts greater than 300,000 IU, which requires ingestion of 120 chewable tablets. Toxicity leads to an acute elevation of intracranial pressure.

(Choice 5) An entire bottle of 100 tablets will need to be ingested to produce fluoride toxicity. Fluoride should not be administered to infants younger than 6 months. From age 6 months to 3 years, a daily supplement of 0.25mg of fluoride is recommended if the water has less than 0.3 parts per million concentration.

239. Question**1 points****Category: Pediatrics**

A 2-year-old boy is rushed to the emergency department by his 21-year-old white mother because he had a sudden-onset nosebleed which has now subsided. He never had any previous episodes. He is "sickly" and suffers from a productive cough and diarrhea that "refuses to go." His stools are greasy and foul-smelling. His appetite is normal, although his diet consists mainly of milk. His mother's boyfriend lives with them and is a chronic alcoholic. He was delivered vaginally without any complications. His weight is at the 25th percentile for his age. Physical examination reveals dry skin and dried blood at the nasal turbinates. What is the most likely cause of this child's failure to thrive?

1. ☐ Lactose intolerance
2. ☐ Parental neglect
3. ☐ Chronic parasitic infection
4. ☐ Constitutional growth delay
5. ☒ Deficiency of pancreatic enzymes ✓
6. ☐ Inadequate dietary intake

INCORRECT ✗**The correct answer is 5.**

Cystic fibrosis is an autosomal recessive disorder that is more common in Caucasian people. The cause is a defect in the CFTR (Cystic fibrosis transmembrane conductance regulator) protein, which leads to impaired chloride transport in tissues and consequent multi-system involvement, particularly:

- (1) Increased salt content in sweat gland secretions
- (2) Thick, viscous secretions in the lungs, pancreas, reproductive tract, and liver.

The patient may have pulmonary manifestations, which begins as a persistent, productive cough, and progresses to obstructive airway disease. A deficiency of pancreatic enzymes leads to fat and protein malabsorption, thereby causing bulky, greasy, foul-smelling stools and subsequent deficiency of fat-soluble vitamins (A, D, E and K). Vitamin A deficiency presents as dry skin, and Vitamin K deficiency presents as epistaxis. Failure to thrive is the result of chronic malabsorption from pancreatic insufficiency. Other possible presentations are meconium ileus at birth and infertility.

(Choice 1) Lactose intolerance is characterized by abdominal cramping, bloating and diarrhea after the intake of lactose-containing products. It does not present with a productive cough.

(Choice 2) Although there should always be a high suspicion for child abuse, parental neglect in this case is a less likely diagnosis. The diarrhea and cough cannot be explained by parental neglect.

(Choice 3) Chronic parasitic infection may be responsible for this patient's diarrhea, but not his cough.

(Choices 4 & 6) Constitutional growth delay and inadequate dietary intake are probable differential diagnoses of this patient's failure to thrive, but these cannot explain the patient's cough and diarrhea.

240. Question

1 points

Category: Pediatrics

A 6-year-old Caucasian male is brought to your office with a two-week history of right shoulder pain. Physical examination reveals localized swelling below the shoulder joint, and x-ray shows a single lytic lesion in the right humeral head. Laboratory analyses show mild hypercalcemia but are otherwise within normal limits. Which of the following is the most likely diagnosis?

1. ☐ Osteoporosis
2. ☐ Sarcoidosis
3. ☒ Langerhans histiocytosis ✓
4. ☐ Primary hyperparathyroidism
5. ☐ Osteogenesis imperfecta

INCORRECT ✗

The correct answer is 3.

The differential for a lytic bone lesion in a child is broad, including infectious (Brodie abscess from osteomyelitis), endocrine (hyperparathyroid osteitis fibrosa cystica), neoplastic (Ewing sarcoma, Langerhans cell histiocytosis, metastases) and idiopathic (benign bone cyst, aneurysmal bone cyst) etiologies. The concomitant hypercalcemia in this patient narrows the differential diagnosis to hyperparathyroid state versus lytic bone neoplasm. Parathyroid adenoma is the most common cause of primary hyperparathyroidism, and typically occurs in patients over fifty years of age. Thus, a lytic bone lesion is most likely. Of the choices listed, only Langerhans cell histiocytosis (LCH, Langerhans cell granulomatosis, histiocytosis X) is known to causes solitary, lytic, long bone lesions. Eosinophilic granuloma, the least severe form of histiocytosis X, generally presents in children and young adults as a solitary bone lesion. This lesion may be painful, have overlying tender swelling, and cause pathological fractures. Though these tumors can be locally destructive, they typically resolve spontaneously and are therefore regarded as benign and treated conservatively.

(Choice 1) Osteoporosis does not cause lytic bone lesions or hypercalcemia.

(Choice 2) Sarcoidosis can cause bone lesions and hypercalcemia, but typically has its onset between ages 20 and 35 years. Sarcoidosis affecting the bone tends to target the hands and feet. There are classically multiple punched out cortical lesions and/or a reticulated pattern of cortical resorption.

(Choice 5) In most cases, osteogenesis imperfecta results from an autosomal dominant defect in Type I collagen. Patients typically present with blue sclerae, diffuse cortical thinning, attenuation of trabeculae and pathological fractures. A solitary lytic bone lesion would be unusual.

241. Question

1 points

Category: Pediatrics

A 3-month-old infant is brought to the emergency department for severe vomiting over the past 6 hours. The mother tells the physician that she has vomited at least 4 times during this period. She also noticed the infant was having difficulty feeding for 2 days. On examination, she is very fussy, and there is a swelling over the left side of the head. CT of the head shows a skull fracture of the left parietal bone with no evidence of intracranial damage. The mother explains that the baby rolled off the sofa onto the floor yesterday. Which of the following is the most appropriate next step in management?

- ☐ Discharge the patient home with instructions concerning post-concussion symptoms
- ☐ Monitor the infant for 12 hours for signs of increased intracranial pressure, discharge the patient home if asymptomatic thereafter
- ☐ Obtain a neurosurgical consultation for the skull fracture
- ☒ Obtain a skeletal survey ✓
- ☐ Repeat the CT scan of the head in 24 hours

INCORRECT ✗

The correct answer is 4.

In this case, the mechanism of the injury (i.e., that the 3-month-old infant rolled off the sofa and hit her head) is inconsistent with the developmental age of the child. A normal 3-month-old infant is unable to roll. Child abuse is suspected in this scenario. In addition, when a parent seeks medical care for the child with a head injury, he or she usually volunteers the mechanism of injury right away, rather than waiting until a CT scan is done to confirm the injury. Such reluctance to give related history is highly suspicious of child abuse. Other clues of injury secondary to child abuse include a changing history, a conflicting history, different caretakers, and a history that is inconsistent with the severity or the type of the injury. In this case, a skeletal survey is indicated to detect any other skeletal injury and to document the findings for legal purpose. A detailed and well-documented physical examination should also be performed to identify any other injury.

Other physical findings that are highly suggestive of child abuse include bruises of buttocks, cigarette burns, circular burns from hot water immersion, human bites, lash marks, spiral fractures of the long bones, subdural hematoma, and multiple bony injuries at different stages of healing. Child abuse continues to be a major problem in this country and is a major threat to the wellness of affected children.

Pediatricians should always be advocates of the children; they are required by law to report any suspected case of child abuse.

(Choice 1) Discharging the patient home is inappropriate because a full evaluation of other possible injuries related to child abuse has not been completed. Failure to recognize child abuse may lead to further serious injury of the victim.

(Choice 2) The problem with 12-hour monitoring of this child is similar to the choice above. Further investigation of the child abuse is necessary. If this is not a child abuse case, monitoring may be useful to detect any development of signs of increased cranial pressure, such as worsening vomiting, lethargy, seizure activities, and Cushing's triad (hypertension, bradycardia, and change in respiration).

(Choice 3) Neurosurgical consultation is not indicated without any evidence of intracranial hemorrhage. A simple linear fracture of the skull does not require immediate surgical intervention.

(Choice 5) Repeating the CT scan in 24 hours is not helpful if the infant remains clinically stable.

242. Question

1 points

Category: Pediatrics

A 1-year-old child is brought in for a well baby checkup. His parents report that he has been of good health and began walking a few weeks earlier. They are concerned that he tends to bump into things and falls more than his older sister did. Family history is significant for retinoblastoma. On examination, the pediatrician notes leukocoria of the left eye. No significant lymphadenopathy is present, and there is no enlargement of the liver or spleen. The child's height and weight are normal for his age. A complete ophthalmologic examination reveals retinoblastoma in the child's left eye, and enucleation is performed. This patient has the highest lifetime risk for which of the following conditions?

1. ☐ Glaucoma
2. ☐ Glioblastoma
3. ☐ Malignant melanoma
4. ☒ Osteosarcoma
5. ☐ Squamous cell carcinoma

INCORRECT ❌

The correct answer is 4.

Retinoblastoma is the most common intraocular malignancy in infants and children. With early diagnosis and treatment, survival is greater than 90%. However, patients with a germline retinoblastoma mutation have a substantial risk (as high as 70%) for having a second high-grade malignancy. The most common secondary non-ocular tumors associated with retinoblastoma is osteosarcoma. Physicians must always have a high index of suspicion of osteosarcoma in these patients when there is a new onset of bone pain or new bony lesions. **(Choice 1)** Infantile or congenital glaucoma is associated with other congenital anomalies, such as aniridia, Sturge-Weber syndrome, Marfan syndrome, and neurofibromatosis. There is, however, no association with retinoblastoma. **(Choices 2, 3 & 5)** Other secondary tumors, such as brain tumors (glioblastoma), malignant melanoma, and squamous cell carcinoma, are also associated with retinoblastoma, but to a much lesser degree than osteosarcoma.

243. Question

1 points

Category: Pediatrics

A 13-year-old girl returns to her physician for followup of a strep throat, for which she had been treated 3 weeks previously. After performing a throat culture, the physician asks how school is going. There is dead silence. Her mother says that her daughter has missed the last 4 weeks of school. Which of the following is the most appropriate initial step in management?

1. ☐ Contract with the girl to go back to school as you explore the problem ✓
2. ☐ Write a medical excuse for her until the throat culture results come back
3. ☐ Tell them you must report her to the school authorities for truancy
4. ☐ Send the mother for supportive counseling
5. ☐ Send the girl for psychotherapy

INCORRECT ✗

The correct answer is 1.

Prolonged absence from school is a serious problem. It is important to sort out the reasons why this teenager is not attending school. If she is staying home with her mother's knowledge, then she is probably being kept home by mother to look after another sibling or to keep her mother company. This school refusal may represent separation anxiety, and there could be underlying conflicts of separation. After her illness with strep throat, this teenager may be having difficulties transitioning back to school. Perhaps she dreads going back to a particular class or teacher. It is important to establish whether her school absence is related to something at home or at school. Importantly, everyone needs to help this teenager return to school, even if a contract needs to be made that she will go only for part of the day.

(Choice 2) The physician should not write a medical excuse to keep her home for any more days. This is not the answer and may only prolong the problem. Many times in such situations families want a doctor's note in support of keeping the child at home and arranging for a home tutor. This should not be done.

(Choice 3) The physician should not report the girl as truant. Truancy is when a student is absent from school but is not at home. Instead the student may be away with peers playing or doing something else. There is no history of truancy in this case. The school is surely aware of the girl's absence from school.

(Choice 4) Sending the mother for counseling is not the best choice. The mother may have underlying separation problems herself. Many mothers of children with school phobia may have a history of depression or have had school phobia themselves during their own childhood. All issues need to be explored over time.

(Choice 5) Sending the girl to psychotherapy seems premature until the physician explores what is going on. At this point, she has not even returned to school. It will take some skill in uncovering the underlying issues and seeing that this teenager gets the help she needs.

244. Question**1 points****Category: Pediatrics**

A 3-month-old Jewish infant is brought to the emergency department because of a generalized seizure 1 hour ago. He is lethargic, weighs 2.7kg (61b), and has a doll-like face with fat cheeks, relatively thin extremities, and a protuberant abdomen. His liver is felt 5cm (2in) below the right costal margin. His kidneys are enlarged. His blood sugar level is 40mg/dl. His serum uric acid, total cholesterol, triglycerides and lactic acid levels are elevated. The levels of his liver transaminases are normal. What is the most likely cause of this infant's symptoms?

1. ☒ Glucose-6-phosphatase deficiency ✓
2. ☐ Acid maltase deficiency
3. ☐ Deficiency of glycogen debranching enzyme activity
4. ☐ Deficiency of branching enzyme activity
5. ☐ Liver phosphorylase deficiency

INCORRECT ✗

The correct answer is 1.

Glucose-6-phosphatase deficiency is also known as type I glycogen storage disease and Von-Gierke's disease. As its name suggests, this condition is caused by deficient glucose-6-phosphatase in the liver, kidneys, and intestinal mucosa. The typical patient, as described in the vignette, is 3-4 months of age with hypoglycemia, lactic acidosis, hyperuricemia, and hyperlipidemia. Hypoglycemic seizures may occur. The characteristic features include a doll-like face (i.e., fat cheeks), thin extremities, short stature, and a protuberant abdomen (due to the enlarged liver and kidneys). The spleen and heart are normal.

(Choice 2) Acid maltase deficiency is also known as type II glycogen storage disease and Pompe's disease. Although patients with this condition also have hepatomegaly, they usually present in the first few weeks of life as a "floppy baby" with feeding difficulties, macroglossia, and heart failure (due to progressive hypertrophic cardiomyopathy).

(Choice 3) Type III glycogen storage disease is caused by a deficiency of glycogen debranching enzyme activity. Similar clinical manifestations (e.g., hepatomegaly, hypoglycemia, hyperlipidemia, and growth retardation) may make it initially difficult to distinguish from type I glycogen storage disease; however, their laboratory findings differ. Patients with glycogen storage III disease have elevated levels of liver transaminases, fasting ketosis, and normal blood lactate and uric acid concentrations. Other pertinent findings are splenomegaly and normal kidneys.

(Choice 4) Deficiency of branching enzyme activity results in type IV glycogen storage disease or amylopectinosis. The typical patient is in his first 18 months of life with hepatosplenomegaly and failure to thrive. The most common pathology is progressive cirrhosis of the liver.

(Choice 5) Patients with liver phosphorylase deficiency have a benign course, and typically present with hepatomegaly and growth retardation early in childhood. Mild hypoglycemia, hyperlipidemia, and hyperketosis may occur. Lactic acid and uric acid levels are normal.

245. Question

1 points

Category: Pediatrics

A 3-day-old girl has trouble feeding and pulmonary congestion. The mother says that the infant is so busy breathing, that she literally has no time to suckle. The girl was born at home, with the delivery attended by a midwife. Physical examination confirms that she is in respiratory distress and shows bounding peripheral pulses with a loud continuous precordial machinery like murmur. X-ray films show increased pulmonary vascular markings. Shortly thereafter, the infant goes into overt heart failure. Which of the following would most likely be required to correct this problem?

1. ☐ Indomethacin
2. ☐ Digitalis and diuretics
3. ☐ Emergency surgical closure of atrial septal defect
4. ☐ Emergency surgical closure of ventricular septal defect
5. ☒ Emergency surgical division of patent ductus arteriosus ✓

INCORRECT ✗

The correct answer is 5.

The diagnosis should be easy to make, since the "machinery-like murmur" is classic for patent ductus arteriosus. In premature infants who are not in failure, closure can be achieved medically with the use of indomethacin. Non-premature infants are less likely to respond, and infants in overt heart failure cannot wait at all and need immediate surgical correction.

(Choice 1), as noted above, is ideal for the premature infant who is not in failure.

(Choice 2) will indeed be used as the infant is rushed to surgery, but, as the question is framed, they alone cannot correct the problem.

(Choices 4 & 5) The infant does not have either an atrial septal defect or a ventricular septal defect. Neither of those gives a continuous machinery-like murmur or produces bounding peripheral pulses.

246. Question

1 points

Category: Pediatrics

A 6-year-old boy has had a fever for 8 days. He just finished a 5-day course of amoxicillin for otitis media. On examination, his temperature is 38.6°C (101.4°F). He has meningismus and palsy of the left sixth cranial nerve. Cerebrospinal fluid (CSF) analysis reveals 200 white cells per mL with 80% lymphocytes and 20% polymorphonuclear leukocytes, glucose of 18 mg/dL, protein of 260 mg/dL, and a negative Gram stain. There is basilar enhancement without focal lesions on CT. Which of the following CSF tests will most likely identify the cause of meningitis?

1. ☐ Antigen test for *Cryptococcus*
2. ☐ Bacterial culture
3. ☒ Culture for mycobacteria ✓
4. ☐ Latex agglutination test for pneumococcus
5. ☐ Test for *Treponema pallidum*

INCORRECT ✗

The correct answer is 3.

Most cases of meningitis in children will be one of two types: bacterial, usually caused by pneumococci or meningococci, or viral, usually caused by enteroviruses. Analysis of the CSF can be very helpful in differentiating the various types of meningitis. Clinicians should also be familiar with other treatable causes of meningitis. Tuberculous meningitis and fungal meningitis have similar clinical presentations and CSF findings. Affected patients have a subacute presentation, focal neurologic abnormalities (in this case a sixth cranial nerve palsy), and a CSF pleocytosis with a predominance of lymphocytes (100–400 cells/mL), a low glucose level (20–45 mg/dL), and an increased protein concentration (100–500 mg/dL). Rapid diagnosis of tuberculous meningitis may be difficult, because most patients will have negative results on acid-fast smear. Results of cerebral CT are usually abnormal, showing enhancement of the cerebral cortex, cerebral edema, infarctions, or obstructive hydrocephalus. Accordingly, the findings in this patient are most consistent with tuberculous meningitis, and mycobacterial culture will be the most helpful CSF test.

(Choice 1) The cryptococcal antigen test has good sensitivity and specificity and has replaced the India ink stain for diagnosis. In approximately 95% of cases of meningitis due to *Coccidioides immitis*, complement fixing antibodies will be present in the CSF.

(Choice 2) is important in the work-up of bacterial meningitis. Acute bacterial meningitis is typically characterized by an elevated CSF white blood cell count with a predominance of neutrophils, an elevated CSF protein concentration, a low CSF glucose level, and positive findings on Gram stain and culture. Prior treatment with oral antibiotics does not alter these diagnostic parameters significantly, although the yield of the CSF Gram stain and culture may be somewhat decreased.

(Choice 4) Latex agglutination tests for pneumococcal or meningococcal antigens rarely are helpful. The CSF Gram stain has a much higher likelihood of providing early diagnostic help.

(Choice 5) *Treponema pallidum* is the cause of syphilis. Clinical manifestations of CNS disease usually appear during the tertiary stages of the disease. A picture of aseptic meningitis can occur rarely and maybe confirmed by a reactive CSF Venereal Disease Research Laboratories (VDRL) test. Classic neuro-syphilis has an indolent course, and psychiatric or neurologic symptoms will not manifest for decades following the initial infection.

247. Question

1 points

Category: Pediatrics

A 6-year-old boy is brought to the ER with a two-day history of difficulty walking. He is dragging his right leg and seems to have weakness in his right arm. He also complains of headaches. The family just emigrated from Eastern Europe. According to his parents, the child suffers from delayed growth compared to his peers and does not like to engage in active play. His lips and fingers turn blue when he cries. His vital signs today are a blood pressure of 100/70 mmHg, pulse of 90/min, temperature of 38.3 ° C (102.0 ° F), and respiratory rate of 22/min. Which of the following is most likely responsible for his current complaints?

1. ☐ Glycogen storage disorder
2. ☐ Demyelinating disorder
3. ☐ Lipid storage disorder
4. ☐ Malignancy
5. ☒ Brain abscess ✓

INCORRECT ✖

The correct answer is 5.

Several elements of this patient's history suggest that he has a background congenital heart disease, including his poor growth, poor energy (manifesting as lack of desire to play with other children), and cyanotic episodes. However, the new-onset headaches, fevers, and focal neurologic changes are concerning for a brain abscess. Children with congenital heart disease, particularly right-to-left shunts, are at increased risk for developing brain abscesses. Brain imaging should be performed as the next step in the workup.

248. Question**1 points****Category: Pediatrics**

A previously healthy 5-month-old boy has been irritable and has had a decreased oral intake for 2 days. His rectal temperature is 37.4 C (99.3 F); pulse is 220/min, and respirations are 50/min. The radial and posterior tibial pulses are diminished with good brachial and femoral pulses. ECG shows tachycardia, QRS complexes are narrow without preceding P waves. Which of the following is the most appropriate initial step in management?

1. ☐ Administer adenosine intravenously
2. ☐ Administer verapamil intravenously
3. ☒ Apply an ice-filled plastic bag to the entire face for 5 to 10 seconds ✓
4. ☐ Cardiac pacing
5. ☐ Perform synchronized direct current cardioversion

INCORRECT ✗

The correct answer is 3.

This child most likely has supraventricular tachycardia. To be more specific, it is atrioventricular re-entry tachycardia (AVRT), a relatively common cardiac emergency among infants. In this condition, the electric impulse is conducted through the AV node, then reenters the atria via an accessory atrioventricular pathway, producing the arrhythmia. The QRS complexes are narrow, and there are no preceding P waves. The pulse is usually 220-270/min. The best means of converting this rhythm to sinus rhythm is to interrupt the re-entry circuit by blocking atrial to ventricular conduction within the AV node. Both vagal maneuvers and drugs that block AV node conduction can be effective treatments. Applying an ice bag to the face is believed to cause a powerful vagal nerve discharge. Most authorities advise using a large plastic bag filled with ice that covers the entire face of the infant; adding some water to the bag appears to achieve the lowest temperature. The face should be covered for only about 5 seconds because the infant will become apneic during the maneuver.

(Choice 1) is an effective and safe method of blocking AV nodal conduction and converting an AVRT to normal sinus rhythm. However, this therapy requires good IV access because the drug must be administered as a rapid bolus. Following this bolus with a rapid infusion of 5-10 mL normal saline ensures that adenosine will be carried through the central circulation before it is metabolized within seconds. Because obtaining IV access can stress these already compromised infants, adenosine should be administered only if a properly performed ice bag maneuver has failed.

(Choice 2) effectively blocks AV conduction, but its use in infants younger than 1 year is extremely dangerous and is contraindicated. Serious side effects include asystole, bradycardia, and shock.

(Choice 4) is the treatment for bradycardia secondary to third degree or complete AV block. It is required in neonates with ventricular rates less than or equal to 50/min who have evidence of heart failure.

(Choice 5) should be reserved for the rare infant who presents in a moribund state, because application of an ice bag to the face often can reinstate sinus rhythm even in infants who appear quite ill.

249. Question

1 points

Category: Pediatrics

A neonate is examined following a protracted breech delivery. One of the infant's arms is partially paralyzed. The affected arm is adducted and internally rotated at the shoulder, and the forearm is pronated. Which of the following is the most likely diagnosis?

1. ☐ Bell palsy
2. ☒ Erb palsy ✓
3. ☐ Klumpke palsy
4. ☐ Pseudobulbar palsy
5. ☐ Supranuclear palsy

INCORRECT ✗

The correct answer is 2.

Brachial plexus injuries can occur in neonates who have difficult deliveries with shoulder dystocia, breech extraction, or hyperabduction of the neck. This infant has Erb palsy, which occurs when the upper part of the brachial plexus is injured and causes the signs illustrated in the question stem. Ipsilateral paralysis of the diaphragm may also be present. Sensory changes are usually not present and, if seen, suggest a much more serious tear or avulsion of the brachial plexus. Fortunately, most cases of Erb palsy resolve within 3 months, and the only treatment needed is a protection of the shoulder from excessive motion by immobilizing the arm across the upper abdomen and daily passive range-of-motion exercises performed gently on involved joints starting at 1 week of age. Cases persisting longer than 3 months may require MRI to evaluate both the extent of the injury and the possibility of surgical repair.

(Choice 1) causes unilateral facial paralysis, typically in adults.

(Choice 3) is due to injury to the lower part of the brachial plexus and causes paralysis of the hand and wrist, often accompanied by ipsilateral Horner syndrome (miosis and ptosis). Like Erb palsy, it can occur during delivery and usually resolves spontaneously in less than 3 months. Passive range-of-motion exercises are the only treatment required.

(Choice 4) is a degenerative disorder of the nervous system that, over a period of years (often starting in adulthood), causes muscle stiffness and weakness in muscles innervated by the lower cranial nerves and thus produces difficulty swallowing.

(Choice 5) is a rare disorder of adults with loss of eye movements, muscular rigidity, and difficulty swallowing.

250. Question

1 points

Category: Pediatrics

A 3-month-old boy is brought to the pediatrician because of a red growth on his arm. The pregnancy had been uncomplicated, and the infant has been meeting all development milestones. He has been healthy so far and has received all scheduled immunizations. He is currently being breast-fed. His skin was clear at birth, but when he was 2 months old, his mother noted a light red growth on his arm. Within the past month, it has increased in size and has turned bright red. Which of the following is the most appropriate treatment for this disorder?

1. ☒ Observation 
2. ☐ Topical corticosteroids
3. ☐ Argon laser therapy
4. ☐ Radiation therapy
5. ☐ Surgery

INCORRECT 

The correct answer is 1.

This child has a strawberry hemangioma, which arises when vascular tissue fails to communicate with adjoining tissue. This enlarges and creates a raised erythematous tumor. Strawberry hemangiomas develop after birth and have a benign course if left alone. They enlarge in the first year of life and then regress. In 90% of patients, the strawberry hemangioma resolves by 9 years of age.

(Choice 2) These lesions are not inflammatory in nature, and topical steroids have not been shown to be of benefit in their treatment.

(Choice 3) If the hemangioma is pressing on a vital organ, argon laser therapy may be needed.

(Choice 4) These lesions are not malignant; therefore, radiation is not needed.

(Choice 5) Surgical resection is rarely needed in these cases.

251. Question

1 points

Category: Pediatrics

A 4-year-old male is brought to the physician with fever and headache. His symptoms began two days ago with low-grade fever, cough, and congestion. Last night, he developed a temperature of 102°F (38.9°C) and became fussy and less active. Today, he is crying and complaining of a headache. His parents report that he has vomited twice today. In the office, his temperature is 102.5°F (39.0°C), pulse is 110/min, and respiratory rate is 20/min. On examination, he is irritable and shows signs of photophobia. His oropharynx is erythematous. Nuchal rigidity is present and when the neck is flexed, the patient flexes his lower extremities. The remainder of the physical examination is normal. Lumbar puncture is performed and the results are shown below.

CSF

Glucose: 60 mg/dL

Protein: 80 mg/dL

RBC: 10/mm³

WBC: 100/mm³

Neutrophils: 10%

Lymphocytes: 70%

Monocytes: 20%

Gram stain: negative

Which of the following organisms is most likely responsible for this patient's presentation?

1. ☐ Streptococcus pneumoniae
2. ☐ Mycobacterium tuberculosis
3. ☐ Epstein-Barr virus
4. ☐ Neisseria meningitidis
5. ☒ Echovirus ✓

INCORRECT ✗

The correct answer is 5.

Viral meningitis is a usually self-limited inflammation of the leptomeninges caused by a viral infection. Almost 90% of cases are caused by non-polio enteroviruses, such as echovirus and coxsackievirus. The incidence of viral meningitis decreases with increasing age. Infants are most commonly affected, and disease morbidity and mortality is highest in this group. Patients with viral meningitis can present with a viral prodrome of constitutional and upper respiratory symptoms with low-grade fever. Over the next 36–48 hours, the patient develops a high fever, headache, irritability, and nuchal rigidity. Focal neurologic signs are not usually seen. The patient may also present with seizures. Other symptoms of enteroviral infection may also be seen such as pharyngitis, rash, or herpangina. In viral meningitis, the cerebrospinal fluid (CSF) will show pleocytosis with lymphocytic predominance, although neutrophils may predominate early in the course. The protein level is normal to slightly elevated and the glucose level is normal. CSF gram stain will not show any organisms. Treatment is supportive; in most patients, symptoms resolve within 7–10 days.

(Choices 1 & 4) Bacterial meningitis caused by *Streptococcus pneumoniae* or *Neisseria meningitidis* will have a similar presentation. However, CSF examination will show an increased cell count with a predominance of neutrophils, increased protein, and decreased glucose. Gram stain will show bacteria.

(Choice 2) CSF in tuberculous meningitis also shows a mildly elevated white cell count with lymphocyte predominance; the protein will be very high and glucose will be low. However, the presentation tends to be subacute rather than acute.

(Choice 3) Viral meningitis can be caused by Epstein-Barr virus, but this is a much less common cause than the enteroviruses.

252. Question

1 points

Category: Pediatrics

A mother brings her 7-year-old son to the clinic because, over the past several days, his urine has become pink and his eyes have looked puffy. About 2 weeks ago, he missed school because of fever and a sore throat. On examination, the boy's blood pressure is 130/85 mm Hg, his eyelids and scrotum appear puffy, and he has 1+ tibial edema. No rashes are noted. Which of the following is the most likely diagnosis?

1. ☒ Acute poststreptococcal glomerulonephritis
2. ☐ Hemolytic-uremic syndrome
3. ☐ Henoch-Schonlein purpura
4. ☐ Nephrotic syndrome
5. ☐ Vesicoureteral reflux

INCORRECT ❌

The correct answer is 1.

Acute poststreptococcal glomerulonephritis is characterized by hematuria with either no or mild edema and elevated blood pressure. In cooler weather, it is often preceded by a sore throat. During warmer weather, a skin infection, such as impetigo, may be the preceding infection. This illness is thought to be mediated by immune complexes, and serum complement (C3) is reduced. When edema is mild, it is most easily seen in loose, thin tissues, such as the eyelids or scrotum. It is uncommon before 3 years of age.

(Choice 2) is a systemic disease, which often follows an acute diarrheal illness and occurs most frequently in children younger than 4 years. Enteropathogenic *Escherichia coli* strains (O157:H7) have been associated with this illness. Following an attack of gastroenteritis, the child becomes pale and lethargic, and urine output falls. Examination often shows edema, petechiae, and hepatosplenomegaly. It is a major cause of renal failure in children.

(Choice 3) is associated with a characteristic petechial or purpuric rash, followed by edema. Some patients have bloody stools and abdominal cramping. Glomerulonephritis may occur 1-2 months after the first symptoms appear and usually presents as asymptomatic hematuria.

(Choice 4) The characteristic clinical manifestation of nephrotic syndrome is edema. In addition, children with nephrotic syndrome have proteinuria, hypoproteinemia, and hyperlipidemia. Although individuals with poststreptococcal glomerulonephritis and Henoch-Schonlein purpura may develop nephrotic syndrome, hematuria is rare in nephrotic syndrome.

(Choice 5) is usually discovered during an evaluation for a urinary tract infection. Significant hematuria or edema is uncommon.

253. Question

1 points

Category: Pediatrics

A 7-year-old boy presents with tenderness and erythema of one knee joint. He has had troubles with infections since about 3 months of age. A brother and a maternal uncle both died of infectious disease at an early age. A detailed immunologic evaluation performed at 2 years of age demonstrated plasma IgG less than 50 mg/100 mL. Normal numbers of circulating T cells and normal cellular immunity were found. The boy had been treated monthly since then with IV immunoglobulin. This therapy had markedly reduced, but not eliminated, the boy's infection rate. Which of the following is the most likely pathogen to cause infectious arthritis in this patient?

1. ☐ Aspergillus
2. ☐ Herpes
3. ☐ Mycobacteria
4. ☒ Mycoplasma
5. ☐ Toxoplasma

INCORRECT ❌

The correct answer is 4.

The immunodeficiency disease described is Bruton (X-linked) hypogammaglobulinemia. This condition is due to a defective B-cell specific tyrosine kinase, coded for by the XLA gene, which facilitates proliferation of pre-B cells in the marrow. The immunologic studies detailed in the question stem are typical of this condition. Affected children are vulnerable to recurrent infections that typically begin at about 3 months of age, when the maternal IgG that crosses the placenta is exhausted. Organisms that are likely to be a particular problem in these antibody-deficient children include Haemophilus, pneumococcus, Mycoplasma, Ureaplasma, Campylobacter, Giardia, and enteroviruses. Septic arthritis is a common problem because there are no antibodies coating and protecting the joint surfaces; common causative organisms include Mycoplasma, Haemophilus, and Pneumococcus. The other organisms listed are important pathogens in patients with depressed cell-mediated immunity.

(Choice 1) The fungus Aspergillus can cause lung infections.

(Choice 2) can cause severe encephalitis.

(Choice 3) can cause lung infections (tuberculosis, atypical mycobacterial infections).

(Choice 5) is a protozoan associated with multisystem infections.

254. Question

1 points

Category: Pediatrics

A woman comes to an emergency department because she is in labor. She has had no prenatal care. Her baby is delivered and appears to be of about 32 weeks' gestation. The newborn is very pale and shows severe, generalized edema. Cord-blood hematocrit is 22%, and cord-blood bilirubin is 7 mg/dL. Ultrasound examination demonstrates pleural effusions, ascites, cardiomegaly, and hepatomegaly. Which of the following is the most likely diagnosis?

1. ☐ ABO incompatibility
2. ☐ Beta thalassemia
3. ☐ Congenital spherocytosis
4. ☐ Sickle cell anemia
5. ☒ Rh incompatibility

INCORRECT ❌

The correct answer is 5.

The infant has hydrops fetalis (erythroblastosis fetalis), which is due to severe hemolytic anemia in the fetus or newborn. Some of these infants die in utero of anemia or during delivery of asphyxiation, since they are so anemic that they cannot tolerate hypoxia. Survivors have many problems, including kernicterus (bilirubin levels high enough to damage the brain), congestive heart failure (high output failure secondary to anemia), hepatosplenomegaly (mostly secondary to extramedullary hematopoiesis), and generalized edema. Although, in theory, hydrops fetalis can occur with any severe intrauterine hemolytic anemia, Rh incompatibility (Rh positive fetus in an Rh negative mother) is the most common cause.

(Choice 1) can cause fetal anemia, but is usually not sufficiently severe to cause the full-blown picture of hydrops fetalis.

(Choices 2 & 4) Beta thalassemia and sickle cell anemia do not usually become clinically evident until about the 6th month of life, when the infant completes the switch from making fetal type hemoglobin to adult type hemoglobin.

(Choice 3) can cause anemia at birth, but is usually not sufficiently severe to cause hydrops fetalis.

255. Question

Category: Pediatrics

1 points

An 11-month-old male infant is brought to the office for a well-baby examination. He was born at term and weighed 2.7 kg (6 lb) at birth. His mother's work is "full time," so he is mostly taken care of by his grandmother. His mother is concerned that he doesn't get to go outdoors since his grandmother has osteoarthritis and her outdoor activity is limited. Physical examination reveals widely open fontanelles and a ping-pong-ball sensation on pressing firmly over the occiput. There are rounded enlargements on the costochondral junctions, and thickening of the wrists and ankles. A horizontal depression is seen along the lower border of the chest. X-ray reveals cupping and fraying of the distal ends of the radius and ulna. What is the most likely diagnosis?

1. ☐ Scurvy
2. ☒ Vitamin D deficiency rickets
3. ☐ Nonrachitic craniotabes
4. ☐ Chondrodystrophy

INCORRECT ❌

The correct answer is 2.

The history, physical and radiologic findings of this infant are diagnostic for rickets. This condition usually presents after several months of being Vitamin D-deficient. The typical patients are low-birth weight infants, un-supplemented dark-skinned infants, infants with inadequate sun exposure, and solely breastfed infants. Common early manifestations are craniotabes (ping-pong ball sensation over the occiput or posterior parietal bones), rachitic rosary (enlargement of the costochondral junctions), and thickening of the wrists and ankles. Other characteristic findings are Harrison groove (horizontal depression on lower border of chest) and a large anterior fontanelle. The diagnosis can be confirmed by obtaining the serum calcitriol level (decreased), alkaline phosphatase level (increased), and observing the characteristic radiologic changes (cupping and fraying of the distal ends of long bones, and double contour along the lateral outline of the radius). Treatment includes oral Vitamin D administration and adequate sunlight (or artificial light) exposure. The current recommendation to prevent rickets is oral Vitamin D supplementation, beginning during the first 2 months of life and continuing until early adolescence.

(Choice 1) Scurvy is commonly seen in 6 to 24-month-old infants with a characteristic history of Vitamin C deficiency. Clinical manifestations are general tenderness, pseudoparalysis (frog position), bluish-purple and spongy/swollen gums over the upper incisors, and depression of the sternum. A rosary similar to rickets may be observed, but the angulation of the beads in patients with scurvy is usually sharper than in those with rickets. The diagnosis is confirmed radiologically (pencil-point thinness of the long bones' cortex, and sharply outlined epiphyseal ends).

(Choice 3) Nonrachitic craniotabes is unlikely in this patient because this condition usually presents in the immediate postnatal period, and disappears before rachitic softening of the skull occurs during the 2nd-4th months of life.

(Choice 4) Patients with chondrodystrophy may also manifest with "rosary"; however, radiologic findings will show irregular, concave outlines of the distal ends of the bones. Chondrodystrophy is therefore a less likely diagnosis in this patient.